Retinal Vasculitis

Primarily

Both and

Primarily

Per Stephen Foster MD: When confronted with a retinal vasculitis, the first step is to determine to which of these categories it belongs.
Per Stephen Foster MD: When confronted with a retinal vasculitis, the first step is to determine to which of these categories it belongs.
Retinal Vasculitis

Retinal Vasculitis

- Primarily Arterial
- Both Arterial and Venous
- Primarily Venous

(Note: The BCSC Retina book asserts that simultaneous involvement of both retinal arterioles and venules is the rule, and that isolated disease of either is distinctly uncommon)
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:

? ? ?

Both Arterial and Venous

Primarily Venous
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- SLE (systemic lupus erythematosus)
- PAN (polyarteritis nodosa)
- Churg-Strauss

Both Arterial and Venous

Primarily Venous
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN
- Churg-Strauss

Both Arterial and Venous

Primarily Venous

Neither the *Retina* nor *Uveitis* book addresses Churg-Strauss, so with all due respect to Dr. Foster, we will not consider it further. Just file it away as a very rare cause of retinal arteritis.
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous

Primarily Venous:
Think of these 3 diseases:
- ?
- ?
- ?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous

Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Toxoplasmosis
- Birdshot
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous

Primarily Venous:
Think of these 1 diseases:
- Sarcoid
- Toxoplasmosis
- Birdshot

The Retina nor Uveitis books only briefly mention the vasculitic component of toxo and birdshot, so with respect again, we will not consider them further. Just remember that vasculitis is a part of these conditions.
Retinal Vasculitis

- Primarily Arterial:
  - Think of these 2 diseases:
    - SLE
    - PAN

- Both Arterial and Venous:
  - Think of these 3 diseases:
    - ?
    - ?
    - ?

- Primarily Venous:
  - Think of these 1 disease:
    - Sarcoid
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet’s
- Wegener’s
- Multiple sclerosis

Primarily Venous:
Think of these 1 disease:
- Sarcoid
Primarily Venous: Think of these 3 diseases:

- Retinal Vasculitis
- PAN
- Behçet’s
- Wegener’s
- Multiple sclerosis

Primarily Arterial: Think of these 2 diseases:

- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:

- PAN
- SLE
- Behçet’s
- Wegener’s
- Multiple sclerosis

Other than mentioning that it is associated with “periphlebitis,” neither the Retina nor Uveitis book addresses MS in the context of vasculitis. Again, file it away.
Primarily Venous:
Think of these 3 diseases:

Primarily Arterial:
Think of these 2 diseases:

Both Arterial and Venous:
Think of these 2 diseases:

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The term ‘Wegener’s granulomatosis’ has fallen out of favor. What term is preferred in its place?
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of this 1 disease:
- Sarcoid

The term ‘Wegener’s granulomatosis’ has fallen out of favor.
What term is preferred in its place?
‘Granulomatosis with polyangiitis’
Primarily Venous: Think of these 3 diseases:

Primarily Arterial: Think of these 2 diseases:

Both Arterial and Venous: Think of these 2 diseases:

- Behçet’s
- Granulomatosis with polyangiitis

The term ‘Wegener’s granulomatosis’ has fallen out of favor.

What is the preferred name? ‘Granulomatosis with polyangiitis’

Why is the name ‘Wegener’s granulomatosis’ no longer preferred?

Retinal Vasculitis

PAN

SLE

Sarcoid
Primarily Venous:
Think of these 3 diseases:

Retinal Vasculitis

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Sarcoid

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

The term ‘Wegener’s granulomatosis’ has fallen out of favor.

Why is the name ‘Wegener’s granulomatosis’ no longer preferred? Because Dr. Wegener was a Nazi
Primarily Venous:
Think of these 3 diseases:
Sarcoid
Retinal Vasculitis
Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
17
SLE
Both Arterial and Venous:
Think of these 3 diseases:
Behçet's
MS
Wegener's
Who is the classic lupus pt?
A woman of childbearing age
Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women
What proportion of lupus pts manifest retinal findings?
About 5-10%
Why is lupus-associated retinal arteritis such an ominous finding?
It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality
Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
With the big dogs: Plasmapheresis + IV cyclophosphamide acutely
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS

Both Arterial and Venous:
Think of these 3 diseases:
- Sarcoid
- Birdshot
- Behçet's

Primarily Venous:
Think of these 3 diseases:
- Wegener's

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Retinal Vasculitis

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Who is the classic lupus pt?
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Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?

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Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
Behçet's
MS (multiple sclerosis)

Primarily Venous:
Think of these 3 diseases:
Toxoplasmosis
Sarcoid
Birdshot
Wegener's

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Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?
--Skin lesions (eg, the classic malar rash)
--Arthralgias

What proportion of pts will manifest ophthalmic findings?
About half

Lupus' ophthalmic manifestations are protean. Which two are most common?
--Lid-skin involvement in a rash--Sjögren syndrome
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN  
SLE  
24

Both Arterial and Venous:
Think of these 3 diseases:
Sarcoid  
Toxoplasmosis  
Birdshot

Primarily Venous:
Think of these 3 diseases:
Behçet’s  
MS (multiple sclerosis)  
Wegener’s

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A woman of childbearing age

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A woman of childbearing age

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What proportion of pts will manifest ophthalmic findings?
About half
Retinal Vasculitis

Primary Arterial: Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Primary Venous: Think of these 3 diseases:
- Sarcoid
- Toxoplasmosis
- Birdshot

Why is lupus-associated retinal arteritis such an ominous finding? It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality.

Who is the classic lupus pt? A woman of childbearing age.

Is there a racial predilection? Yes, black and Hispanic women are at greater risk than are white women.

What proportion of lupus pts manifest retinal findings? About 5-10%.

Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?
- Skin lesions (eg, the classic malar rash)
- Arthralgias

What proportion of pts will manifest ophthalmic findings? About half.

Lupus’ ophthalmic manifestations are protean. Which two are most common?
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- ...
Who is the classic lupus pt?
A woman of childbearing age

Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?
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Lupus’ ophthalmic manifestations are protean. Which two are most common?
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--Sjögren syndrome
Retinal Vasculitis

**Primarily Arterial:**
Think of these 3 diseases:
- PAN
- SLE
- Behçet's

**Primarily Venous:**
Think of these 3 diseases:
- Sarcoid
- Birdshot
- MS

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?

Why is lupus-associated retinal arteritis such an ominous finding?
It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality

Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
With the big dogs: Plasmapheresis + IV cyclophosphamide acutely
Retinal Vasculitis

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

Why is lupus-associated retinal arteritis such an ominous finding?
It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality

Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
With the big dogs: Plasmapheresis + IV cyclophosphamide acutely
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Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

This range represents a compromise between inconsistencies in the BCSC series:
--Rate of lupus retinopathy per the Retina book: 3-10%
--Rate per the Uveitis book: 3-29%
(I rounded to 30 to make it easier to remember)
Retinal Vasculitis

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

What retinal findings may occur?
- Occlusive events: Asymptomatic cotton wool spots
- If unlucky, an infarcted macula

Why is lupus-associated retinal arteritis such an ominous finding?
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What proportion of lupus pts manifest retinal findings?
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What retinal findings may occur?
Occlusive events: Asymptomatic cotton wool spots if ‘lucky,’ an infarcted macula if not
Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

*CWS are the classic manifestation of lupus retinopathy!*

Occlusive events: Asymptomatic cotton wool spots, if ‘lucky,’ an infarcted macula if not

(No question—proceed when ready)
Retinal Vasculitis

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

_CWS are the classic manifestation of lupus retinopathy!_

Occlusive events: Asymptomatic cotton wool spots if ‘lucky,’ an infarcted macula if not

Is macular infarction also a classic manifestation of lupus retinopathy?
Retinal Vasculitis

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic cotton wool spots if ‘lucky,’ an infarcted macula if not

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare
Who is the classic lupus pt?
A woman of childbearing age

What is the fundamental difference between the pathophysiology of CWS and macular infarction?

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic if ‘lucky,’ an infarcted macula if not.

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare.
Who is the classic lupus pt?
A woman of childbearing age

What is the fundamental difference between the pathophysiology of CWS and macular infarction?
It’s the size of the vessel involved. CWS result from infarction of tiny end-arterioles of the arterial tree, whereas macular infarction involves the relatively ginormous central retinal artery, or an immediate branch thereof.

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic
if ‘lucky,’ an infarcted macula

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare
Who is the classic lupus pt? A woman of childbearing age

What is the fundamental difference between the pathophysiology of CWS and macular infarction? It’s the size of the vessel involved. CWS result from infarction of tiny end-arterioles of the arterial tree, whereas macular infarction involves the relatively ginormous central retinal artery, or an immediate branch thereof.

How does this explain the fact that CWS are vastly more common than macular infarction?

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic if ‘lucky,’ an infarced macula if not.

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Retinal Vasculitis

Who is the classic lupus pt?
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What is the fundamental difference between the pathophysiology of CWS and macular infarction?
It’s the size of the vessel involved. CWS result from infarction of tiny end-arterioles of the arterial tree, whereas macular infarction involves the relatively ginormous central retinal artery, or an immediate branch thereof.

How does this explain the fact that CWS are vastly more common than macular infarction?
It’s because lupus is a microvasculopathy, and thus much more likely to affect end-arterioles.

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic, if ‘lucky,’ an infarcted macula, not.

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare.
Who is the classic lupus pt?
A woman of childbearing age

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Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

Why is lupus-associated retinal arteritis an ominous finding?
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What proportion of lupus pts manifest retinal findings?
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Why is lupus-associated retinal arteritis an ominous finding?
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**Retinal Vasculitis**

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What proportion of lupus pts manifest retinal findings?
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Why is lupus-associated retinal arteritis an ominous finding?
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Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
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With the big dogs: Plasmapheresis + IV cyclophosphamide acutely
Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?

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In a nutshell, what is the pathophysiology of PAN?
Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?

Subacute episodes of focal necrotizing inflammation of arteries

PAN is strongly associated with seropositivity for what virus?

Hepatitis B

Who is the typical PAN pt?

A male between 40 and 60 years old

Is there a racial predilection?

No

Primarily Arterial

Think of these 3 diseases:

PAN

SLE

Both Arterial and Venous

Think of these 3 diseases:

Sarcoid

Behçet's

MS (multiple sclerosis)

Wegener's

Primarily Venous

Think of these 3 diseases:

Sarcoid

Behçet's

MS (multiple sclerosis)
Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they?

Classic PAN affects medium and small arteries, whereas MP affects smaller arteries.
Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases: PAN, SLE, MS

Both Arterial and Venous: Think of these 3 diseases: Behçet’s, Wegener’s

Primarily Venous: Think of these 3 diseases: Sarcoid, Birdshot

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they? ‘Classic’ PAN, and microscopic polyangiitis (MP)

Who is the typical PAN pt?
A male between 40 and 60 years old

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Hepatitis B
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet’s
- Wegener’s
- Sarcoid

Primarily Venous:
Think of these 3 diseases:
- Birdshot

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Subacute episodes of focal necrotizing inflammation of arteries

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What is the difference between the two?
Retinal Vasculitis

**In a nutshell, what is the pathophysiology of PAN?**
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they?

'Classic' PAN, and microscopic polyangiitis (MP)

What is the difference between the two? Classic PAN affects medium and small arteries, whereas MP affects smaller arteries.
In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they?
'Classic' PAN, and microscopic polyangiitis (MP)

What is the difference between the two?
Classic PAN affects medium and small arteries, whereas MP affects smaller arteries

What’s the difference between a ‘small’ artery and a ‘smaller’ artery?
Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they?
'Classic' PAN, and microscopic polyangiitis (MP)

What is the difference between the two?
Classic PAN affects medium and small arteries, whereas MP affects smaller arteries

What's the difference between a 'small' artery and a 'smaller' artery?
Rule of thumb: Classic PAN only affects arteries large enough to be named, whereas MP only affects vessels smaller than that

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE

Both Arterial and Venous:
Think of these 3 diseases:

Primarily Venous:
Think of these 3 diseases:
Sarcoid
Birdshot
Behçet's
MS
Wegener's
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
Behçet's

Primarily Venous:
Think of these 3 diseases:
MS
Wegener's

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Is it a common, or uncommon condition?
Uncommon

Who is the typical PAN pt?
A male between 40 and 60 years old

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Hepatitis B
Retinal Vasculitis

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SLE
PAN

Primarily Arterial
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Primarily Venous
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Behçet's
Sarcoid
MS (multiple sclerosis)

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A male between 40 and 60 years old

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
Behçet’s

Primarily Venous:
Think of these 3 diseases:
MS
Wegener’s

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Is it a common, or uncommon condition?
Uncommon

Who is the typical PAN pt?
A male between 40 and 60 years old

Is there a racial predilection?
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Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
Behçet's

Primarily Venous:
Think of these 3 diseases:
Sarcoid
MS
Wegener's

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

What is the typical presentation of PAN? (Note: It's not ocular.)
Constitutional symptoms: Fever, fatigue, weight loss. Renal vasculitis resulting in secondary HTN is common.

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Hepatitis B
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
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Constitutional symptoms: Fever, fatigue, weight loss. Renal vasculitis resulting in secondary HTN is common.

What proportion of PAN pts develop ocular involvement?
About 20%
Retinal Vasculitis

Primarily Arterial:
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About 20%

What is the 5 year survival rate for untreated PAN?
A miserable 10%

What is the 5 year survival rate of treated PAN?
A robust 80%
Retinal Vasculitis

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What is the typical ocular presentation of PAN? (Head's up: It's probably not what you think.)
Peripheral ulcerative keratitis (PUK). It can cause scleritis (both anterior and posterior) as well.

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
Behçet's

Both Arterial and Venous:
Think of these 3 diseases:
Sarcoid
Wegener's
MS (multiple sclerosis)

Primarily Venous:
Think of these 3 diseases:
Hepatitis B
PAN
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OK fine. What is the typical posterior-pole presentation of PAN?
In a manner consistent with severe HTN
Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Is it a common, or uncommon condition?
Uncommon

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No

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Advance to the next slide to see what this entails…

In a manner consistent with severe HTN
What process mediates damage caused by acute, severe HTN?
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of the retinal arterioles
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

?  ?  ?
Retinal Vasculitis

Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

Retinopathy  Choroidopathy  Optic Neuropathy
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm

Of the retinal arterioles

What categories of pathology can result?

Retinopathy

Choroidopathy

Optic Neuropathy
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

Retinopathy
Choroidopathy
Optic Neuropathy

Cotton-wool spots
Retinal hemorrhages
Macular star
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

Retinopathy
- Cotton-wool spots
- Retinal hemorrhages
- Macular star

Choroidopathy
- Leads to...

Optic Neuropathy
- Leads to...
- ?
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

Retinopathy

Cotton-wool spots
Retinal hemorrhages
Macular star

Choroidopathy

Leads to...

Choroidal infarction

Leads to...

Exudative RD

Optic Neuropathy

Retinal Vasculitis
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

Retinopathy
- Cotton-wool spots
- Retinal hemorrhages
- Macular star

Choroidopathy
- Leads to...
- Choroidal infarction
- Leads to...
- Exudative RD

Optic Neuropathy
- Leads to...

Retinal Vasculitis
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm
Of the retinal arterioles

What categories of pathology can result?

Retinopathy  Choroidopathy  Optic Neuropathy

Cotton-wool spots  Retinal hemorrhages  Macular star

Leads to…

Choroidal infarction

Leads to…

Exudative RD

Disc edema, hemorrhages
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases: Ischamia, Retinal Vasculitis
  - SLE
  - PAN

- Both Arterial and Venous: Think of these 2 diseases: Behçet’s, Granulomatosis

- Primarily Venous: Think of this 1 disease: Sarcoid

Who is the typical BD pt?
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous: Think of this 1 disease:
- Sarcoid

Who is the typical BD pt? An individual of ‘Silk Road’ descent age 25-35
Retinal Vasculitis

- **Primarily Arterial**: Think of these 2 diseases:
  - SLE
  - PAN

- **Both Arterial and Venous**: Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

- **Primarily Venous**: Think of this 1 disease:
  - Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?

An ancient trading route extending from the eastern Mediterranean to the Far East

- The Middle East; Turkey; China; Korea; Japan
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous: Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East

What regions/countries are found along the Silk Road?
Retinal Vasculitis

Primarily Arterial:
- Think of these 2 diseases:
  - SLE
  - PAN

Both Arterial and Venous:
- Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

Primarily Venous:
- Think of this 1 disease:
  - Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East

What regions/countries are found along the Silk Road?
The Middle East; Turkey; China; Korea; Japan
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- SLE
- PAN
- Behçet’s

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Granulomatosis

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?

Which of these areas has the highest rate of BD?
The Middle East; Turkey; China; Korea; Japan
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous: Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?

Which of these areas has the highest rate of BD?
Turkey

What regions/countries are found along the Silk Road?
The Middle East, Turkey, China, Korea, Japan
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous: Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- PAN
- SLE

Retinal Vasculitis

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Wegener’s
- Granulomatosis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers
Retinal Vasculitis

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet's
- Sarcoid
- Granulomatosis

An individual of ‘Silk Road’ descent age 25-35

What is the classic nonocular complaint in BD?
Recurrent oral ulcers

Are the oral ulcers painful?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrence oral ulcers

Are the oral ulcers painful?
Yes, very
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - SLE
  - PAN

- Both Arterial and Venous: Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

- Primarily Venous: Think of this 1 disease:
  - Sarcoid

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
--Skin rash
--Genital ulcers
--Arthralgias
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
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Primarily Venous:
Think of this 1 disease:
- Sarcoid

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What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
- Genital ulcers
- Arthralgias

What is the classic rash?
Erythema nodosum

The classic location:
The pre-tibial region
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - SLE
  - PAN
- Both Arterial and Venous: Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis
- Primarily Venous: Think of this 1 disease:
  - Sarcoid

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Recurrent oral ulcers

In addition to... What is the classic rash?
Skin rash
- Erythema nodosum
- Genital ulcers
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In addition to... What is the classic rash?
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- Erythema nodosum
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Retinal Vasculitis

Primarily Arterial:
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Primarily Venous:
Think of this 1 disease:
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What is the classic rash?
Erythema nodosum

Where is the classic location?
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
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Both Arterial and Venous: Think of these 2 diseases:
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The pre-tibial region
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Primarily Venous:
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What the classic nonocular complaint in BD?
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What is the cause of BD?
It is unknown at this time
Retinal Vasculitis

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Think of these 2 diseases:
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- PAN

Both Arterial and Venous:
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- Both Arterial and Venous:
  - Think of these 2 diseases:
    - Behçet's
    - Granulomatosis

- Primarily Venous:
  - Think of this 1 disease:
    - Sarcoid

**Is ocular involvement common in BD?**

- Yes; estimates of ocular involvement run as high as 70%

**Does BD pose a significant threat to long-term ocular health, and/or vision?**

- Yes, severe vision loss results in as many as 25% of ocular BD cases

**Who is the typical BD pt?**

- An individual of 'Silk Road' descent age 25-35

**What is the classic nonocular complaint in BD?**

- Recurrent oral ulcers

**In addition to oral ulcers, what three other nonocular findings are common?**

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Granulomatosis Retinal Vasculitis

Retinal Vasculitis

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- Mos def--severe vision loss results in as many as 25% of ocular BD cases
Retinal Vasculitis

Primarily Arterial:
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- SLE
- PAN

Primarily Venous:
Think of this 1 disease:

- Sarcoid

Both Arterial and Venous:
Think of these 2 diseases:

- Behçet’s
- Granulomatosis

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)

Does BD pose a significant threat to long-term ocular health, and/or vision?
Mos def--severe vision loss results in as many as 25% of ocular BD cases

In addition to oral ulcers, what three other nonocular findings are common?
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**Retinal Vasculitis**

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Think of these **2** diseases:
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- Granulomatosis

**Primarily Venous:**
Think of this **1** disease:
- Sarcoid

**Is ocular involvement common in BD?**
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**Does BD pose a significant threat to long-term ocular health, and/or vision?**
Mos def--severe vision loss results in as many as 25% of ocular BD cases

**In addition to oral ulcers, what three other nonocular findings are common?**
- Skin rash
- Genital ulcers
- Arthralgias

**What is the most common form of ocular involvement? (It’s not retinal vasculitis.)**
Acute nongranulomatous anterior uveitis, often with a hypopyon
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- Behçet’s

Primarily Venous:
Think of these 2 diseases:
- Panarteritis nodosa
- Sarcoid

Both Arterial and Venous:
Think of these 2 diseases:
- Wegener’s
- Retinal Vasculitis

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
Acute nongranulomatous anterior uveitis, often with a hypopyon.

What are three other nonocular findings common in BD?
- Skin rash
- Genital ulcers
- Arthralgias

Does BD pose a significant threat to long-term ocular health, and/or vision?
Mos def--severe vision loss results in as many as 25% of ocular BD cases.

In addition to oral ulcers, what three other nonocular findings are common?
--Skin rash
--Genital ulcers
--Arthralgias

Is ocular involvement common in BD?
Yes; estimates of ocular involvement run as high as 70%.

When does ocular BD typically occur?
Most often when BD is systemic, often within 4 years of its onset.
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Wegener's

Primarily Venous:
Think of this 1 disease:
- Granulomatosis Sarcoid

Is ocular involvement common in BD?

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
Acute nongranulomatous anterior uveitis, often with a hypopyon

Does BD pose a significant threat to long-term ocular health, and/or vision?
Most def—severe vision loss results in as many as 25% of ocular BD cases

In addition to oral ulcers, what three other nonocular findings are common?

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:

- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:

- Behçet’s
- Granulomatosis

Primarily Venous:
Think of this 1 disease:

- Sarcoid

Is ocular involvement common in BD?

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Does BD pose a significant threat to long-term ocular health, and/or vision?

- Mos def--severe vision loss results in as many as 25% of ocular BD cases

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)

- Acute nongranulomatous anterior uveitis, often with a hypopyon

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?

- Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and a significant cause of ocular morbidity and vision loss
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

What is ocular involvement common in BD?
- Is ocular involvement common in BD?
- What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
- Acute nongranulomatous anterior uveitis, often with a hypopyon

How is the diagnosis of BD made?

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
- Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and a significant cause of ocular morbidity and vision loss
Retinal Vasculitis

Both **Arterial** and **Venous**:
- Think of these **2** diseases:
  - Behçet's
  - Granulomatosis

Primarily **Arterial**:
- Think of these **2** diseases:
  - SLE
  - PAN

Primarily **Venous**:
- Think of this **1** disease:
  - Sarcoid

Is ocular involvement common in BD?
- Yes; estimates of ocular involvement run as high as 70%

Does BD pose a significant threat to long-term ocular health, and/or vision?
- Mos def--severe vision loss results in as many as 25% of ocular BD cases

What is the most common form of ocular involvement? (It's not retinal vasculitis.)
- Acute nongranulomatous anterior uveitis, often with a hypopyon

How is the diagnosis of BD made?
- It is a clinical diagnosis--there are no specific tests for it

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
- Yes--it is an occlusive vasculitis, so ischemia is a common occurrence, and a significant cause of ocular morbidity and vision loss
Granulomatosis Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

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How is the diagnosis of BD made?
It is a clinical diagnosis—there are no specific tests for it.

What is the HLA association for BD?
There are several; HLA-B51 is associated with ocular BD.

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and a significant cause of ocular morbidity and vision loss.
**Retinal Vasculitis**

**Primarily Arterial:**
Think of these 2 diseases:
- SLE (Sjögren's Syndrome Lupus Erythematosus)
- PAN (Polyarteritis Nodosa)

**Both Arterial and Venous:**
Think of these 2 diseases:
- Behçet's
- Granulomatosis

**Primarily Venous:**
Think of this 1 disease:
- Sarcoid

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**What is the most common form of ocular involvement in BD?**

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Primarily Arterial: Think of these 2 diseases:
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Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of this 1 disease:
- Sarcoid

What is the classic triad of granulomatosis with polyangiitis (GPA)?

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Retinal Vasculitis

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- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous:
Think of this 1 disease:
- Sarcoid

What is the classic triad of granulomatosis with polyangiitis (GPA)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
--small and medium-sized arteries and veins
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:

Both Arterial and Venous: Think of these 2 diseases:

What is the classic triad of granulomatosis with polyangiitis (GPA)? Necrotizing vasculitis of:
-- the upper and lower respiratory tract
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-- small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement?

Primarily Venous: Think of this 1 disease:

Behçet's
Granulomatosis with polyangiitis
Sarcoid
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:

Primarily Venous:
Think of these 2 diseases:

Both Arterial and Venous:
Think of these 2 diseases:

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic triad of granulomatosis with polyangiitis (GPA)?
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Behçet's

Granulomatosis with polyangiitis

Sarcoid
Retinal Vasculitis

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Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:

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- Behçet's

Primarily Venous: Think of this 1 disease:

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- the upper respiratory tract
- the lower respiratory tract
- the kidneys
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What is the classic presentation of lower respiratory tract involvement?
- Cough (+/- hemoptysis), wheezing, etc
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Think of these 3 diseases:

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--the kidneys
--small and medium-sized arteries and veins

Primarily Venous:
Think of this 1 disease:

- Sarcoid

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

Granulomatosis with polyangiitis
Retinal Vasculitis

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What specific pathologic renal condition is caused by GPA?

Glomerulonephritis

Sinusitis with bloody discharge

A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc
Retinal Vasculitis

Primarily Arterial:
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SLE
Behçet’s

Primarily Venous:
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What specific pathologic renal condition is caused by GPA?
Glomerulonephritis
Retinal Vasculitis

Of the three, which does the Uveitis book describe as “the most characteristic clinical feature” of GPA?

Primarily Arterial:
Think of these 3 diseases:

What is the classic presentation of upper respiratory tract involvement?
Sinusitis

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

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--the kidneys
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Primarily Venous:
Think of this 1 disease:

Sarcoid

What specific pathologic renal condition is caused by GPA?
Glomerulonephritis

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Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
Granulomatosis with polyangiitis

Behçet’s

What is the classic triad of granulomatosis with polyangiitis (GPA)?
Necrotizing vasculitis of:
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Glomerulonephritis
Retinal Vasculitis

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Sinus dz

Primarily Arterial:
Think of these 3 diseases:

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Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

Granulomatosis with polyangiitis

Primarily Venous:
Think of this 1 disease:

Sarcoid

What is the classic triad of granulomatosis with polyangiitis (GPA)?
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--the upper and lower respiratory tract
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What specific pathologic renal condition is caused by GPA?
Glomerulonephritis
What is the classic finding on CT? 
Arterial: 
Think of these 2 diseases: 
What is the classic presentation of upper respiratory tract involvement? 
Sinusitis with bloody discharge 
What is the classic presentation of lower respiratory tract involvement? A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc 

Primarily Venous: 
Think of this 1 disease: 

What is the classic triad of granulomatosis with polyangiitis (GPA)? 
Necrotizing vasculitis of: 
--the upper and lower respiratory tract 
--the kidneys 
--small and medium-sized arteries and veins 

What specific pathologic renal condition is caused by GPA? 
Glomerulonephritis 

Retinal Vasculitis 

Retinal Vasculitis
retinal vasculitis

arterial:
think of these 3 diseases:

what is the classic presentation of upper respiratory tract involvement?
sinusitis with bloody discharge

what is the classic presentation of lower respiratory tract involvement?
variety of pulmonary s/s: cough (+/- hemoptysis), wheezing, etc

what is the classic finding on ct?
bony destruction of the sinuses

behavior's

granulomatosis with polyangiitis

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necrotizing vasculitis of:
-- the upper and lower respiratory tract
-- the kidneys
-- small and medium-sized arteries and veins

what specific pathologic renal condition is caused by gpa?
glomerulonephritis

venous:
think of this 1 disease:
sarcoid

primarily arterial:

sle

pan

primarily venous:

behrer's disease
Retinal Vasculitis

What is the classic finding on CT?
Bony destruction of the sinuses

Arterial:
Think of these 2 diseases:

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

- Behçet’s
- Granulomatosis with polyangiitis

What is the classic triad of granulomatosis with polyangiitis (GPA)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract
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What specific pathologic renal condition is caused by GPA?
Glomerulonephritis

Primarily Venous:
Think of this 1 disease:
Sarcoid
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:

PAN
SLE
Behçet’s

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic finding on CT?
Bony destruction of the sinuses

Retinal

What is the classic finding on CT?
Cavitary lung lesions

Arterial:
Think of these 2 diseases:

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Sinusitis with bloody discharge

Granulomatosis with polyangiitis

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Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
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What specific pathologic renal condition is caused by GPA?
Glomerulonephritis

Primarily Venous:
Think of this 1 disease:
Sarcoid

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What specific pathologic renal condition is caused by GPA?
Glomerulonephritis
Retinal Vasculitis

**Arterial:**
Think of these diseases:
- PAN
- SLE
- Behçet's
Both

**Think of these 3 diseases:**
- PAN
- SLE
- Behçet's

**Primarily Arterial:**
Think of these 3 diseases:
- PAN
- SLE
- Behçet's

**Primarily Venous:**
Think of these 1 disease:
- Sarcoid

**What is the classic triad of granulomatosis with polyangiitis (GPA)?**
Necrotizing vasculitis of:
--the **upper** and **lower** respiratory tract
--the **kidneys**
--small and medium-sized arteries and veins

**What specific pathologic renal condition is caused by GPA?**
Glomerulonephritis

**What is the classic finding on CT?**
Cavitary lung lesions

**What is the classic finding on CT?**
Bony destruction of the sinuses

**What is the classic presentation of lower respiratory tract involvement?**
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

- Behçet's

**What lab findings result?**
Hematuria and proteinuria

- Granulomatosis with polyangiitis

**What is the classic presentation of upper respiratory tract involvement?**
Sinusitis with bloody discharge
Retinal Vasculitis

What is the classic finding on CT?
Cavitary lung lesions

What is the classic presentation of lower respiratory tract involvement?
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Retinal

Arterial:
Think of these 3 diseases:
- PAN
- SLE
- Behçet’s

Venous:
Think of this 1 disease:
- Sarcoid

Primarily Arterial:

Primarily Venous:

What is the classic finding on CT?
Bony destruction of the sinuses

Granulomatosis with polyangiitis
Retinal Vasculitis

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What lab findings result?
Hematuria and proteinuria

What specific pathologic renal condition is caused by GPA?
Glomerulonephritis

Two other manifestations are common as well—what are they?
- --
- --

Granulomatosis with polyangiitis

What is the classic triad of granulomatosis with polyangiitis (GPA)?
Necrotizing vasculitis of:
- the upper respiratory tract
- the kidneys

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What specific pathologic renal condition is caused by GPA?
Glomerulonephritis

Hematuria and proteinuria
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Both Arterial and Venous:
Think of these 3 diseases:
- Granulomatosis with polyangiitis
- Sarcoid

Primarily Venous:
Think of this 1 disease:
- Sarcoid

What is the classic finding on CT?
- Bony destruction of the sinuses
- Cavitary lung lesions

What is the classic presentation of upper respiratory tract involvement?
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What is the classic triad of granulomatosis with polyangiitis (GPA)?
- Necrotizing vasculitis of:
  - the upper and lower respiratory tract
  - the kidneys

Two other manifestations are common as well—what are they?
- Rash
- Arthralgias

What lab findings result?
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What specific pathologic renal condition is caused by GPA?
- Glomerulonephritis
What proportion of GPA pts have ophthalmic involvement?

What is the classic triad of granulomatosis with polyangiitis (GPA)?
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Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
Behçet’s

Both Arterial and Venous:
Think of these 3 diseases:
2

Primarily Venous:
Think of these 3 diseases:
Sarcoid
Granulomatosis with polyangiitis

What proportion of GPA pts have ophthalmic involvement?

About half

Orbital inflammation

What is the next most common manifestation? (Still not retinal.)
Scleritis (including PUK)
**Retinal Vasculitis**

What proportion of GPA pts have ophthalmic involvement? About half

What is the most common manifestation of that involvement? (It’s not retinal.) Orbital inflammation

What is the next most common manifestation? (Still not retinal.) Scleritis (including PUK)

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Bony destruction of the sinuses

What is the classic finding on CT?

Orbital inflammation

Scleritis (including PUK)

Hematuria and proteinuria

Rash

Arthralgias

Granulomatosis with polyangiitis

Glomerulonephritis

Necrotizing vasculitis of:
--- the upper and lower respiratory tract
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Retinal Vasculitis
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Bony destruction of the sinuses
Cavitary lung lesions

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Retinal Vasculitis

OK then, what proportion of GPA pts manifest vasculitis?
About 10%

What is the classic triad of granulomatosis with polyangiitis (GPA)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys

Two other manifestations are common as well—what are they?
--Rash
--Arthralgias

What is the classic finding on CT?
Bony destruction of the sinuses

What is the classic finding on CT?
Cavitary lung lesions

What lab findings result?
Hematuria and proteinuria

What specific pathologic renal condition is caused by GPA?
Glomerulonephritis

What is the classic presentation of upper respiratory tract involvement?
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As CWS, or arterial occlusions (branch or central)

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**What is the classic triad of granulomatosis with polyangiitis (GPA)?**
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--- small and medium-sized arteries and veins

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**What is the prognosis for untreated GPA?**
Dismal—the 1-yr mortality rate is 80%

**What is the treatment of choice?**
Steroids + cyclophosphamide

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**What is the next most common manifestation of GPA?**
Scleritis (including PUK)

**What proportion of GPA pts have ophthalmic involvement?**
About half

**What is the most common manifestation of that involvement?**
(It’s not retinal.) Orbital inflammation

**Orbital inflammation**

**OK then, what proportion of GPA pts manifest vasculitis?**
About 10%

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--the upper and lower respiratory tract
--the kidneys

What is the treatment of choice?
Steroids + cyclophosphamide monotherapy?

Cyclophosphamide is an extremely harsh drug—why not tx with steroids alone?

Steroid monotherapy?

Granulomatosis with polyangiitis

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Cyclophosphamide is an extremely harsh drug—why not tx with steroids alone? 
Because steroid monotherapy for GPA is associated with an increased risk of mortality

Steroid monotherapy? No!
In broadest terms, how would you describe sarcoid?

Primarily Venous: Think of these diseases:

Sarcoid
Retinal Vasculitis

In broadest terms, how would you describe sarcoid?
It is a multi-organ disease characterized by the presence of noncaseating granulomas.

In which organ/system does sarcoid most commonly involve?
The lungs.

Which other organ systems can be involved?
Pretty much all of them.

What percentage of sarcoid pts have ocular involvement?
About 50%.

Who is the typical sarcoid pt in the US?
An African-American.

During what age-range does dz onset typically occur?
Usually 20 to 50.
Retinal Vasculitis

In broadest terms, how would you describe sarcoid? It is a multi-organ disease characterized by the presence of noncaseating granulomas.

Primarily Venous: Think of these 3 diseases:

- Wegener’s
- Sarcoid
- Behçet’s

Primarily Arterial: Think of these 3 diseases:

- PAN
- SLE
- PRP

Primarily Venous: Think of these 3 diseases:

- Sarcoid
- PAN
- SLE

In the US, the typical sarcoid pt is an African-American. During what age-range does dz onset typically occur? Usually 20 to 50.
In broadest terms, how would you describe sarcoid?
It is a multi-organ disease characterized by the presence of noncaseating granulomas

What is the cause?

As of this writing, it is unknown

What organ/system is most commonly involved in sarcoid?
The lungs

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Pretty much all of them

What percentage of sarcoid pts have ocular involvement?
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Think of these 3 diseases:

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**How about world-wide? Which ethnic group has the highest prevalence of sarcoid?**
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During what age-range does dz onset typically occur?
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What is the single-best screening test for sarcoid?
Chest X-ray

What is the classic CXR finding?
Bilateral hilar adenopathy

If CXR is read as normal, but clinical suspicion for sarcoidosis remains high, what other imaging modality should be considered?
Thin cut CT of the chest

What is the only way to definitively diagnose sarcoidosis?
Via biopsy demonstrating the presence of noncaseating granulomas
Retinal Vasculitis

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Bilateral hilar adenopathy

If CXR is read as normal, but clinical suspicion for sarcoidosis remains high, what other imaging modality should be considered?
Thin cut CT of the chest

What is the only way to definitively diagnose sarcoidosis?
Via biopsy demonstrating the presence of noncaseating granulomas
Retinal Vasculitis

In broadest terms, how would you describe sarcoid?
It is a multi-organ disease characterized by the presence of noncaseating granulomas.

What is the cause?
As of this writing, it is unknown.

What organ/system is most commonly involved in sarcoid?
The lungs.

Which other organ/systems can be involved?
Pretty much all of them.

What percentage of sarcoid pts have ocular involvement?
About 50.

Who is the typical sarcoid pt in the US?
An African-American.

During what age-range does dz onset typically occur?
Usually 20 to 50.

What is the single-best screening test for sarcoid?
Chest X-ray.

What is the classic CXR finding?
Bilateral hilar adenopathy.

If CXR is read as normal, but clinical suspicion for sarcoidosis remains high, what other imaging modality should be considered?
Thin cut CT of the chest.

What is the only way to definitively diagnose sarcoidosis?
Via biopsy demonstrating the presence of noncaseating granulomas.
Retinal Vasculitis

- Primarily Arterial: Think of these 3 diseases: SLE, PAN
- Both Arterial and Venous: Think of these 2 diseases: Behçet’s, Granulomatosis with polyangiitis
- Primarily Venous: Think of these 1 disease: Sarcoid

What percentage of sarcoid uveitis pts have posterior manifestations?

About 20%
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of these 1 disease:
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What are the two most common posterior manifestations?
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Think of these 1 diseases:
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What percentage of sarcoid uveitis pts have posterior manifestations? About 20

What are the two most common posterior manifestations?
--Granulomas of the choroid, retina and/or optic nerve
--Retinal vasculitis
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
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Think of these 3 diseases:
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What percentage of sarcoid uveitis pts have posterior manifestations?
About 20%

What are the two most common posterior manifestations?
- Granulomas of the choroid, retina, and/or optic nerve
- Retinal vasculitis

Is the vasculitis primarily an arteritis, a phlebitis, or both?
Sarcoid tends to cause a phlebitis (more specifically, a periphlebitis).

What is the classic appearance of sarcoid periphlebitis?
Small yellowish-white nodular granulomas accumulate along venules in a pattern referred to as candle-wax drippings.

What French term for candle-wax drippings is encountered frequently in the ophthalmic literature?
Taches de bougie.
Retinal Vasculitis

Primarily Arterial

Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Both Arterial and Venous

Think of these 3 diseases:
- Wegener's
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What are the two most common posterior manifestations?

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- Retinal vasculitis
What is the first-line med for sarcoid uveitis?

Steroids (Topical, periocular or systemic)

What other treatment is important to include, and why?

Cycloplegia (if there is a significant anterior component), both for comfort and to prevent synechiae

If steroid therapy fails to yield adequate control, what is the next step?

Immunomodulatory therapy (eg, methotrexate, azathioprine or cyclosporine)

What about biologics—are they useful in controlling sarcoid uveitis?

Yes, specifically with infliximab (Remicade)
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Think of these 3 diseases:
PAN
SLE
Behçet’s
Both Arterial and Venous:
Think of these 3 diseases:
Wegener’s
Retinal Vasculitis
Primarily Venous:
Think of these 3 diseases:
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- SLE
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Primarily Arterial and Venous:

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- Retinal Vasculitis
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Retinal Vasculitis

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Primarily Venous: Think of these 1 disease: Sarcoid

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What about biologics--are they useful in controlling sarcoid uveitis?
Yes, specifically with infliximab (Remicade)
Two more conditions worth discussing (by way of a hint: both names are eponyms)
Retinal Vasculitis

- **Primarily Arterial:**
  - Think of these 2 diseases:
    - SLE
    - PAN
    - Susac syndrome

- **Both Arterial and Venous:**
  - Think of these 2 diseases:
    - Behçet’s
    - Granulomatosis with polyangiitis

- **Primarily Venous:**
  - Think of these 1 diseases:
    - Sarcoid
    - Eales disease

**Two more conditions worth discussing (by way of a hint: both names are eponyms)**
Granulomatosis with polyangiitis
Retinal Vasculitis

Both Arterial and Venous:
Think of these 3 diseases:
- SLE
- Sarcoid
- Toxoplasmosis

Primarily Arterial:
Think of these 3 diseases:
- Behçet's
- Granulomatosis with polyangiitis
- Susac syndrome

Primarily Venous:
Think of these 3 diseases:
- Susac syndrome
- Eales disease
- Venous retinal vasculitis

What is Susac syndrome?

- An obliterative retinal vasculopathy
- Primarily Venous
- Bilaterality
- Young-adult female
- Classic retinal finding:
  - Multiple BRAOs occurring at non-branch points
- Classic association:
  - Sensorineural hearing loss

What is Eales disease?

- An obliterative retinal vasculopathy
- Primarily Venous
- Bilaterality
- Classic association:
  - Tests positive for TB
Retinal Vasculitis

Granulomatosis with polyangiitis
Behçet's

Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
SLE
PAN
Sarcoid

Primarily Venous:
Think of these 3 diseases:
Primarily Venous:
Toxoplasmosis

What is Eales disease?
An obliterative retinal vasculopathy
Does it present unilaterally, or bilaterally?
Bilaterally
Who is the classic patient?
Young-adult male from India
What is the classic retinal finding?
Peripheral neovascularization
Classic association?
Tests positive for TB

What is Susac syndrome?
An obliterative retinal vasculitis
Does it present unilaterally, or bilaterally?
Bilaterally
Who is the classic patient?
Young-adult female
What is the classic retinal finding?
Multiple BRAOs occurring at non-branch points
Classic association?
Sensorineural hearing loss

Susac syndrome

Eales disease
What is Susac syndrome?
An obliterative retinal vasculitis

What is Eales disease?
An obliterative retinal vasculitis

Susac syndrome

Eales disease

Retinal Vasculitis
Granulomatosis with polyangiitis (GPA) and Behçet’s disease are both associated with retinal vasculitis.

Primarily Arterial: SLE, PAN

Primarily Venous: Sarcoid, Toxoplasmosis

Both Arterial and Venous: Think of these 3 diseases:

- Susac syndrome
- Eales disease
- Retinal Vasculitis

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- Bilaterally
- Who is the classic patient?
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- What is the classic retinal finding?
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- Does it present unilaterally, or bilaterally?
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- Who is the classic patient?
- Young-adult female
- What is the classic retinal finding?
- Multiple BRAOs occurring at non-branch points
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Retinal Vasculitis

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An obliterative retinal vasculitis

Does it present unilaterally, or bilaterally?

Susac syndrome

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Susac syndrome

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Retinal Vasculitis

Behçet's disease

Granulomatosis with polyangiitis

Retinal Vasculitis

Both Arterial and Venous

Think of these 3 diseases:

SLE
PAN
Sarcoid
Toxoplasmosis

Primarily Arterial

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Think of these 3 diseases:

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Primarily Venous:
Think of these 3 diseases:

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**Retinal Vasculitis**

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- **Who:**

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- **What is Eales disease?**
  - An obliterative retinal vasculitis
- **Does it present unilaterally, or bilaterally?**
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- **Who is the classic patient?**
  - Young-adult female
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  - Multiple BRAOs occurring at non-branch points
- **Classic association?**
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**What is Susac syndrome?**
- An obliterative retinal vasculitis
- **Does it present unilaterally, or bilaterally?**
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- **Who is the classic patient?**
  - Young adult females
Granulomatosis with polyangiitis (GPA) and Behçet’s disease are two of these 2 diseases.

Retinal Vasculitis

- **Susac syndrome**
  - Who: Young adult females

- **Eales disease**
  - Who: Young adult males from India

- **What is Susac syndrome?**
  - An obliterative retinal vasculitis

- **Does it present unilaterally, or bilaterally?**
  - Bilaterally

- **Who is the classic patient?**
  - Young-adult female

- **What is Eales disease?**
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  - Bilaterally

- **Who is the classic patient?**
  - Young-adult female

- **What is the classic retinal finding?**
  - Multiple BRAOs occurring at non-branch points

- **Classic association?**
  - Sensorineural hearing loss
Granulomatosis with polyangiitis

Behçet’s

Retinal Vasculitis

Both Arterial and Venous: Think of these 3 diseases:

- SLE
- Sarcoid
- Toxoplasmosis

Primarily Arterial: Think of these 3 diseases:

Primarily Venous: Think of these 3 diseases:

Who: Young adult males

What is Eales disease?
An obliterative retinal vasculitis

Does it present unilaterally, or bilaterally?
Bilaterally

Who is the classic patient?
Young-adult male from India

What is the classic retinal finding?
Peripheral neovascularization

Classic association?
Tests positive for TB

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Does it present unilaterally, or bilaterally?
Bilaterally

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Young-adult female

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Classic association?
Sensorineural hearing loss

Susac syndrome
Who: Young adult females

Eales disease
Who:
**Retinal Vasculitis**

**Susac syndrome**
*Who: Young adult females*

**Eales disease**
*Who: Young adult males*

**What is Susac syndrome?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult female

**What is Eales disease?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult male from India

**What is Eales disease?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult male from India
Retinal Vasculitis

**Susac syndrome**
*Who:* Young adult **females**
*What:*

**Eales disease**
*Who:* Young adult **males**

**What is Susac syndrome?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult female

**What is the classic retinal finding?**

**What is Eales disease?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
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Young-adult male from India

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**Behçet’s**

**2**

**Retinal Vasculitis**

Both

Arterial and Venous:

Think of these **3** diseases:

- SLE
- PAN
- Sarcoid

**Primarily Arterial:**

Think of these **3** diseases:

- Toxoplasmosis

**Primarily Venous:**

Think of these **3** diseases:

- Susac syndrome
- Eales disease

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**Who:** Young adult **males**

**What:** Peripheral neo

**Associated with:** TB positivity

---

**Who:** Young adult **females**

**What:** Multiple BRAOs

**What is Eales disease?**

An obliterative retinal vasculitis

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Retinal Vasculitis

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Think of these 3 diseases:

- **SLE**
- **Sarcoid**
- **Toxoplasmosis**

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Think of these 3 diseases:

- **Sarcoid**
- **Toxoplasmosis**
- **SLE**

**Primarily Venous:**

Think of these 3 diseases:

- **Susac syndrome**
- **Eales disease**
- **Retinal Vasculitis**

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**Susac syndrome**

Who: Young adult **females**

What: **Multiple BRAOs**

**Eales disease**

Who: Young adult **males**

What: **Peripheral neo**

Associated with: **TB positivity**

Who: Young adult **males**

What: **Peripheral neo**

Associated with: **TB positivity**

**What is Eales disease?**

An obliterative retinal vasculitis

Does it present unilaterally, or bilaterally?

Bilaterally

Who is the classic patient?

Young-adult male from India

What is the classic retinal finding?

Peripheral neovascularization

Classic association?

Tests positive for TB

**What is Susac syndrome?**

An obliterative retinal vasculitis

Does it present unilaterally, or bilaterally?

Bilaterally

Who is the classic patient?

Young-adult female

What is the classic retinal finding?

Multiple BRAOs occurring at non-branch points

Classic association?

Sensorineural hearing loss

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Susac syndrome
Who: Young adult females
What: Multiple BRAOs

Eales disease
Who: Young adult males
What: Peripheral neovascularization
Retinal Vasculitis

**Susac syndrome**
- **Who:** Young adult females
- **What:** Multiple BRAOs
- **Associated with:**

**Eales disease**
- **Who:** Young adult males
- **What:** Peripheral

**What is Susac syndrome?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult female

**What is the classic retinal finding?**
Multiple BRAOs occurring at non-branch points

**Classic associations (Note: there are two)?**

**What is Eales disease?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult male from India

**What is the classic retinal finding?**
Peripheral neovascularization

**Classic association?**
Tests positive for TB

An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult male from India

**What is the classic retinal finding?**
Peripheral neovascularization

**Classic association?**
Tests positive for TB

- **Granulomatosis with polyangiitis**
- **Behçet's disease**
- **Retinal Vasculitis**

- **Both Arterial and Venous:**
  - Think of these 3 diseases:
  - **SLE**
  - **Sarcoid**
  - **Toxoplasmosis**

- **Primarily Arterial:**
  - Think of these 3 diseases:
  - **SLE**
  - **Sarcoid**
  - **Toxoplasmosis**

- **Primarily Venous:**
  - Think of these 3 diseases:
  - **SLE**
  - **Sarcoid**
  - **Toxoplasmosis**

**What is Eales disease?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult male from India

**What is the classic retinal finding?**
Peripheral neovascularization

**Classic association?**
Tests positive for TB
Retinal Vasculitis

**Susac syndrome**
- **Who:** Young adult females
- **What:** Multiple BRAOs
- **Associated with:** Hearing loss; encephalopathy

**Eales disease**
- **Who:** Young adult males
- **What:** Peripheral neo

**What is Susac syndrome?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult female

**What is the classic retinal finding?**
Multiple BRAOs occurring at non-branch points

**Classic associations (Note: there are two)?**
Sensorineural hearing loss; encephalopathy

**What is Eales disease?**
An obliterative retinal vasculitis

**Does it present unilaterally, or bilaterally?**
Bilaterally

**Who is the classic patient?**
Young-adult male from India

**What is the classic retinal finding?**
Peripheral neovascularization

**Classic association?**
Tests positive for TB

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**Granulomatosis with polyangiitis**

**Behçet’s**

**Retinal Vasculitis**

Both Arterial and Venous:
Think of these 3 diseases:

- SLE
- Sarcoid
- Toxoplasmosis

Primarily Arterial:
Think of these 3 diseases:

- Think of these 3 diseases:
- Think of these 3 diseases:

Primarily Venous:
Think of these 3 diseases:
Granulomatosis with polyangiitis

Behçet's

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Retinal Vasculitis

Both Arterial and Venous:

Think of these 3 diseases:

SLE
Sarcoid
Toxoplasmosis

Primarily Arterial:

Think of these 3 diseases:

Primarily Venous:

Think of these 3 diseases:

Who: Young adult males
What: Peripheral neo
Associated with:

Eales disease

Who: Young adult males
What: Peripheral neo
Associated with:

Susac syndrome

Who: Young adult females
What: Multiple BRAOs
Associated with: Hearing loss; encephalopathy

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Multiple BRAOs occurring at non-branch points

Classic associations (Note: there are two)?
Sensorineural hearing loss; encephalopathy
**Retinal Vasculitis**

- **Susac syndrome**
  - **Who:** Young adult females
  - **What:** Multiple BRAOs
  - **Associated with:** Hearing loss; encephalopathy

- **Eales disease**
  - **Who:** Young adult males
  - **What:** Peripheral neo
  - **Associated with:** TB positivity

- **What is Eales disease?**
  - An obliterative retinal vasculitis

  - **Does it present unilaterally, or bilaterally?**
    - Bilaterally

  - **Who is the classic patient?**
    - Young-adult male from India

  - **What is the classic retinal finding?**
    - Peripheral neovascularization

  - **Classic association?**
    - Tests positive for TB

- **What is Susac syndrome?**
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  - **Who is the classic patient?**
    - Young-adult female

  - **What is the classic retinal finding?**
    - Multiple BRAOs occurring at non-branch points

  - **Classic associations (Note: there are two)?**
    - Sensorineural hearing loss; encephalopathy

  - **What is Eales disease?**
    - An obliterative retinal vasculitis

    - **Does it present unilaterally, or bilaterally?**
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    - **Who is the classic patient?**
      - Young-adult male from India

    - **What is the classic retinal finding?**
      - Peripheral neovascularization

    - **Classic association?**
      - Tests positive for TB
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of these 1 disease:
- Sarcoid

Susac syndrome
Who: Young adult females
What: Multiple BRAOs
Associated with: Hearing loss; encephalopathy

Eales disease
Who: Young adult males
What: Peripheral neo
Associated with: TB positivity

High-yield points to remember about these two conditions