In a nutshell, what is scleritis?
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An inflammatory condition characterized by painful infiltrative scleral edema
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Is it serious, or more of a nuisance?
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Very serious--it can be extremely painful, can lead to blindness and loss of the eye, and can be associated with systemic conditions that are potentially lethal
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Is there an age predilection?
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Is there a gender predilection?
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Yes, men are less likely to be affected than women
Scleritis

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Yes, men are less likely to be affected than women
Scleritis

Very basic anatomic division

?  ?  ?
Scleritis

Very basic anatomic division

Anterior  Scleritis  Posterior
Scleritis

- Anterior
- Posterior

? ? ?
Scleritis

- Anterior
  - Diffuse
  - Nodular
  - Necrotizing
- Posterior
Scleritis

- Anterior
  - Diffuse
  - Nodular
  - Necrotizing

- Posterior

?
Scleritis

- Anterior
  - Diffuse
  - Nodular
  - Necrotizing
    - w/ inflammation
    - w/o inflammation

- Posterior
What are three classic signs of anterior scleritis?

--?
--?
--?
What are three classic signs of anterior scleritis?

- Scleral edema
- Sclera has a violaceous hue
- Flamed vasculature has a criss-cross pattern
Scleritis

What are three classic signs of anterior scleritis?
--Scleral edema
--?
--?
Scleral edema in nodular scleritis
What are three classic signs of anterior scleritis?
--Scleral edema
--Sclera has a violaceous hue
--?
What are three classic signs of anterior scleritis?

--Scleral **edema**

--Sclera has a **violaceous** hue

--?
Scleritis

Violaceous hue in diffuse scleritis
What are three classic signs of anterior scleritis?
--Scleral edema
--Sclera has a violaceous hue
--Inflamed vasculature has a criss-cross pattern
What are three classic signs of anterior scleritis?
--Scleral edema
--Sclera has a violaceous hue
--Inflamed vasculature has a criss-cross pattern
Criss-cross deep vasculature in scleritis (look for it until you see it)
Scleritis

- Anterior
  - Diffuse
  - Nodular
  - Necrotizing

- Posterior

Is the nodule of nodular scleritis mobile?

No
Scleritis

Anterior

Diffuse
Nodular

Posterior

Necrotizing

Is the nodule of nodular scleritis mobile?
No

/o inflammation
If a patient has a necrotizing scleritis w/ inflammation, it’s a virtual lock that what sort of disease is the culprit?
If a patient has a necrotizing scleritis w/ inflammation, it's a virtual lock that what sort of disease is the culprit?
A vasculitis
Scleritis

Necrotizing scleritis with inflammation
Scleritis

If a patient has a necrotizing scleritis w/ inflammation, it's a virtual lock that what sort of disease is the culprit?

A 

vasculitis

Is the vasculitis always systemic?
If a patient has a necrotizing scleritis with inflammation, it’s a virtual lock that what sort of disease is the culprit? A vasculitis.

Is the vasculitis always systemic?
No, but it certainly can be.
Scleritis

Scleritis

Anterior

Posterior

Diffuse

Nodular

Necrotizing

w/ inflammation

w/ inflammation

If a patient has a necrotizing scleritis w/ inflammation, what sort of disease is the culprit?
A vasculitis

Is the vasculitis always systemic?
No, but it certainly can be

Here is where the eye dentist* can save someone’s life. The discovery of an ocular vasculitic process should prompt a thorough investigation into whether the ocular disease is a manifestation of a systemic process.

*Eye dentist (slang) Derisive nickname for an ophthalmologist
If a patient has a necrotizing scleritis with inflammation, what sort of disease is the culprit?
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Is the vasculitis always systemic?
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Why is this so important?
- The discovery of an ocular vasculitic process should prompt a thorough investigation into whether the ocular disease is a manifestation of a systemic process.

Here is where the eye dentist can save someone's life.
Scleritis

Anterior

- Diffuse
- Nodular
- Necrotizing
  - w/ inflammation
  - w/o inflammation

Posterior

If a patient has a necrotizing scleritis w/ inflammation, it's a virtual lock that what sort of disease is the culprit?

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Is the vasculitis always systemic?

No, but it certainly can be

Here is where the eye dentist can save someone’s life.

The discovery of an ocular vasculitic process should prompt a thorough investigation into whether the ocular disease is a manifestation of a systemic process.

Why is this so important?

The presence of ocular vasculitis in systemic dz portends a dismal prognosis for the pt. Five-year mortality for such pts approaches 50%!
By what other name is necrotizing scleritis w/o inflammation known?

Scleritis

Anterior

Diffuse  Nodular  Necrotizing

w/ inflammation  w/o inflammation

Is it in fact a noninflammatory disease, as the name implies?
No; inflammation is present on biopsy.

With what systemic disease is it most commonly associated?
Rheumatoid arthritis

How does its presentation differ from that of its 'w/ inflammation' cousin?
Scleromalacia perforans is typically painless, and the eye is not inflamed.
By what other name is necrotizing scleritis w/o inflammation known? Scleromalacia perforans
Scleritis

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Scleritis

Anterior

Diffuse  Nodular  Necrotizing

w/ inflammation  w/o inflammation
Scleritis

Scleritis

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Scleritis

Anterior

Diffuse

Nodular

Necrotizing

w/ inflammation

w/o inflammation
Scleritis

Anterior

Diffuse Nodular Necrotizing

w/ inflammation w/o inflammation

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By what other name is necrotizing scleritis w/o inflammation known? Scleromalacia perforans

Is it in fact a noninflammatory disease, as the name implies? No; inflammation is present on biopsy

With what systemic disease is it most commonly associated? Rheumatoid arthritis

Does scleromalacia perforans tend to occur early, or late in the RA dz course? It tends to occur late, in longstanding cases.
**Scleritis**

- Anterior
  - Diffuse
  - Nodular
  - Necrotizing
    - w/ inflammation
    - w/o inflammation

**By what other name is necrotizing scleritis w/o inflammation known?**
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**Does scleromalacia perforans tend to occur early, or late in the RA dz course?**
It tends to occur late, in longstanding cases
Scleritis

**Anterior**

- Diffuse
- Nodular
- Necrotizing

**Necrotizing**

- **w/ inflammation**
- **w/o inflammation**

---

*By what other name is necrotizing scleritis w/o inflammation known?*

Scleromalacia perforans

*Is it in fact a noninflammatory disease, as the name implies?*

No; inflammation is present on biopsy

*With what systemic disease is it most commonly associated?*

Rheumatoid arthritis

*How does its presentation differ from that of its ‘w/ inflammation’ cousin?*
Scleritis

**By what other name is necrotizing scleritis w/o inflammation known?**
Scleromalacia perforans

**Is it in fact a noninflammatory disease, as the name implies?**
No; inflammation is present on biopsy

**With what systemic disease is it most commonly associated?**
Rheumatoid arthritis

**How does its presentation differ from that of its ‘w/ inflammation’ cousin?**
Scleromalacia perforans is typically painless, and the eye is not inflamed
Scleritis

Necrotizing scleritis w/o inflammation
Is this scleromalacia perforans?
Scleritis

No--this is blue sclera in **Ehlers-Danlos**
Scleritis

Is this scleromalacia perforans?
No--this is blue sclera in OI
Scleritis

Is this scleromalacia perforans?
Scleritis

No--this is **ocular melanocytosis**
Is this scleromalacia perforans?
No--this is oculodermal melanocytosis (aka Nevus of Ota)
From what related disease must scleritis be differentiated?

Scleritis

Anterior

Diffuse

Nodular

w/ inflammation

w/o inflammation
From what related disease must scleritis be differentiated? Episcleritis

Scleritis

- Anterior
  - Diffuse
  - Nodular
    - w/ inflammation
    - w/o inflammation

Episcleritis
From what related disease must scleritis be differentiated? Episcleritis

What is the hallmark feature that separates the two?

Scleritis

- Anterior
  - Diffuse
  - Nodular
    - w/ inflammation
    - w/o inflammation
From what related disease must scleritis be differentiated?
Episcleritis

What is the hallmark feature that separates the two?
Scleritis is painful—the eye hurts, and is usually quite tender.
In contrast, episcleritis is not painful (other than perhaps a mild foreign-body sensation), and the globe is not tender.
From what related disease must scleritis be differentiated?
Episcleritis

What is the hallmark feature that separates the two?
Scleritis is **painful**—the eye hurts, and is usually quite tender.
In contrast, episcleritis is **not** painful (other than perhaps a mild foreign-body sensation), and the globe is **not** tender.

Necrotizing scleritis w/o inflammation is a notable exception to this rule.
What proportion of scleritis patients have an identifiable systemic condition?
What proportion of scleritis patients have an identifiable systemic condition? About 1/2
Scleritis

What proportion of scleritis patients have an identifiable systemic condition?
About 1/2

More specifically, what proportion of Diffuse vs Nodular vs Necrotizing pts have an identifiable systemic condition?

Diffuse ≈ ?
Nodular ≈ ?
Necrotizing ≈ ?
What proportion of scleritis patients have an identifiable systemic condition? About 1/2

More specifically, what proportion of Diffuse vs Nodular vs Necrotizing pts have an identifiable systemic condition? 1/3, 1/3 and 2/3, respectively
What proportion of scleritis patients have an identifiable systemic condition? About 1/2

(2 most common classes?)

½ w/ systemic condition
½ no systemic condition
What proportion of scleritis patients have an identifiable systemic condition? About 1/2

Scleritis

Anterior

- Diffuse
  - w/ inflammation
  - w/o inflammation

- Nodular

- Necrotizing

Posterior

½ w/ systemic condition

½ no systemic condition

CT Disease

Vasculitis

(CT = connective tissue)
What proportion of scleritis patients have an identifiable systemic condition? About 1/2

(3 most common CT diseases?)
Scleritis

What proportion of scleritis patients have an identifiable systemic condition? About 1/2

½ w/ systemic condition
½ no systemic condition

CT Disease
- RA (rheumatoid arthritis)
- SLE (systemic lupus erythematosus)
- SNSAs (seronegative spondyloarthropathies)

Vasculitis
Scleritis

What proportion of scleritis patients have an identifiable systemic condition? About 1/2

½ no systemic condition
½ w/ systemic condition

CT Disease Vasculitis

Scleritis

Anterior

Diffuse
Nodular
Necrotizing

Posterior

To what does the term seronegative spondyloarthropathies refer?
It is an umbrella term for a group of related rheumatologic conditions

To the fact that the rheumatoid factor test is negative in these conditions

Name three common seronegative spondyloarthropathies.
-- Ankylosing spondylitis
-- Reactive arthritis
-- Psoriatic arthritis

Scleritis

½ w/ systemic condition
½ no systemic condition

Scleritis

T Disease Vasculitis

RA (rheumatoid arthritis)
SLE (systemic lupus erythematosus)
SNSAs (seronegative spondyloarthropathies)
Scleritis

What proportion of scleritis patients have an identifiable systemic condition?
About 1/2

½ no systemic condition
½ w/ systemic condition

CT Disease Vasculitis

Anterior Posterior

Diffuse Nodular Necrotizing

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To what does the word seronegative refer?
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SNSAs (seronegative spondyloarthropathies)
Scleritis

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½ no systemic condition
½ w/ systemic condition

Scleritis

Anterior

Diffuse
Nodular
Necrotizing

Posterior

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Scleritis

CT Disease Vasculitis

RA (rheumatoid arthritis)
SLE (systemic lupus erythematosus)
SNSAs (seronegative spondyloarthropathies)

T Disease Vasculitis

1/2 w/ systemic condition
1/2 no systemic condition

RA (rheumatoid arthritis)
SLE (systemic lupus erythematosus)
SNSAs (seronegative spondyloarthropathies)
Scleritis

What proportion of scleritis patients have an identifiable systemic condition?
About 1/2

½ no systemic condition
½ w/ systemic condition

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CT Disease Vasculitis

Anterior

Posterior

Diffuse

Nodular

Necrotizing
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Scleritis

Anterior

Posterior

Diffuse
Nodular
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CT Disease Vasculitis

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Name three common seronegative spondyloarthropathies.
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--?
--?
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What proportion of scleritis patients have an identifiable systemic condition? About 1/2

½ w/ systemic condition
½ no systemic condition

CT Disease
RA
SLE
SNSAs

Vasculitis
?

(3 most common *vasculitides*)
Scleritis

What proportion of scleritis patients have an identifiable systemic condition? About 1/2

½ w/ systemic condition
½ no systemic condition

CT Disease
- RA
- SLE
- SNSAs

Vasculitis
- Wegener’s
- PAN (polyarteritis nodosa)
- GCA (giant-cell arteritis)
What proportion of scleritis patients have an identifiable systemic condition?

About 1/2

½ no systemic condition
½ w/ systemic condition

Which of these disease are associated with episcleritis?

All of them! The DDx for episcleritis is the same as that for scleritis. The difference between these conditions concerns disease probability—the likelihood of an underlying condition is far lower in episcleritis.
Which of these diseases are associated with episcleritis? All of them! The DDx for episcleritis is the same as that for scleritis. The difference between these conditions concerns disease probability—the likelihood of an underlying condition is far lower in episcleritis.
What proportion of scleritis patients have an identifiable systemic condition?

About 1/2

What important-but-often-overlooked source of local scleritis has yet to be mentioned?

Infectious scleritis

A review by Foster and colleagues revealed what infection rate among scleritis pts?

About 7%—yikes! Make sure you keep infectious processes in the back of your mind when working up scleritis patients.
Scleritis

What important-but-often-overlooked source of local scleritis has yet to be mentioned? **Infectious** scleritis

About 1/2 scleritis patients have an identifiable systemic condition?
Scleritis

What proportion of scleritis patients have an identifiable systemic condition?

About 1/2

Anterior Posterior

Diffuse Nodular Necrotizing

w/ inflammation w/o inflammation

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What is the most common event leading to the development of infectious scleritis?

Pterygium surgery (other surgeries, and trauma, are common inciting incidents as well)

What bug is most commonly implicated?

Pseudomonas

How should infectious scleritis be treated?

With both antibiotics and surgical debridement
Scleritis

What proportion of scleritis patients have an identifiable systemic condition?
About 1/2

Scleritis

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Nodular

w/ inflammation

w/o inflammation
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What bug is most commonly implicated? *Pseudomonas*

How should infectious scleritis be treated? With both antibiotics and **surgical debridement**.

**Why surgical debridement? That is, why aren’t abx enough?** Because the sclera is essentially avascular, it is difficult to achieve adequate abx concentrations.
What proportion of scleritis patients have an identifiable systemic condition? About 1/2.

What important but often overlooked source of local scleritis has yet to be mentioned? Infectious scleritis.

A review by Foster and colleagues revealed what infection rate among scleritis patients? About 7%. Make sure you keep infectious processes in the back of your mind when working up scleritis patients!

What is the most common event leading to the development of infectious scleritis? Pterygium surgery (other surgeries, and trauma, are common inciting incidents as well).

What bug is most commonly implicated? Pseudomonas.

How should infectious scleritis be treated? With both antibiotics and surgical debridement.

Why surgical debridement? That is, why aren’t abx enough? The sclera is essentially avascular, so it is difficult to achieve adequate abx concentrations within it.
What signs/symptoms of anterior scleritis do not typically appear in isolated posterior scleritis?
--?
--?
What signs/symptoms of anterior scleritis do not typically appear in isolated posterior scleritis?
--The eye is not red
--No nodules are present

What signs/symptoms of posterior scleritis do not typically occur in anterior scleritis?

Is posterior scleritis more likely or less likely than anterior uveitis to be associated with a systemic condition?
Less likely
Scleritis

Anterior

- Diffuse
- Nodular
- Necrotizing w/ inflammation

Posterior

What signs/symptoms of anterior scleritis do not typically appear in isolated posterior scleritis?

-- The eye is not red
-- No nodules are present

What signs/symptoms of posterior scleritis do not typically occur in anterior scleritis?

-- ?
-- ?
-- ?
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Scleritis

What signs/symptoms of anterior scleritis do not typically appear in isolated posterior scleritis?
--The eye is not red
--No nodules are present

What signs/symptoms of posterior scleritis do not typically occur in anterior scleritis?
--Proptosis
--Disc edema
--Motility disorders
--Retinal/choroidal findings
Scleritis

Posterior scleritis OD: Optic nerve edema
Scleritis

Posterior scleritis: Scleral thickening
Scleritis

Anterior

- Diffuse
- Nodular
- Nodular w/ inflammation

Posterior

- Proptosis
- Disc edema
- Motility disorders
- Retinal/choroidal findings

What signs/symptoms of anterior scleritis do not typically appear in isolated posterior scleritis?

-- The eye is not red
-- No nodules are present

What signs/symptoms of posterior scleritis do not typically occur in anterior scleritis?

Is posterior scleritis more likely or less likely than anterior scleritis to be associated with a systemic condition?
What signs/symptoms of anterior scleritis do not typically appear in isolated posterior scleritis?
--The eye is not red
--No nodules are present

What signs/symptoms of posterior scleritis do not typically occur in anterior scleritis?
--Proptosis
--Disc edema
--Motility disorders
--Retinal/choroidal findings

Is posterior scleritis more likely or less likely than anterior scleritis to be associated with a systemic condition?
Less likely
Scleritis

What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?

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--?
What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?

- The pain radiates to the brow
- The pain is aggravated by ocular rotations
- The pain awakens the pt at night

Scleritis

Anterior

Posterior

w/ inflammation

w/o inflammation
What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?

-- The pain radiates to the **brow**
--?
--?
What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?

--The pain radiates to the **brow**

--The pain is aggravated by ___

--?
What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?

-- The pain radiates to the **brow**

-- The pain is aggravated by **eye movements**

-- ?

w/ inflammation  w/o inflammation
What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?
--The pain radiates to the **brow**
--The pain is aggravated by **eye movements**
--The pain... **five words**
What three characteristics of the pain are a tipoff that you’re dealing with posterior scleritis?

--The pain radiates to the **brow**
--The pain is aggravated by **eye movements**
--The pain **awakens the pt at night**
Scleritis

Anterior
- Diffuse
- Nodular
  - w/ inflammation

Posterior

What easy-to-obtain imaging study can cinch the diagnosis?

- b-scan ultrasonography
  - Choroidal thickening
  - Sub-Tenon's edema

When sub-Tenon's edema involves the space around the optic nerve, what b-scan finding will result?

The classic T sign.
Scleritis

Anterior

Diffuse

Nodular

Posterior

What easy-to-obtain imaging study can cinch the diagnosis? 
b-scan ultrasonography

w/ inflammation
Scleritis

Anterior

Diffuse

Nodular

Posterior

With inflammation w/o inflammation

What easy-to-obtain imaging study can cinch the diagnosis?

b-scan ultrasonography

What will b-scan reveal?

--?

--?
Scleritis

Anterior

- Diffuse
- Nodular
  - w/ inflammation

Posterior

- W/ inflammation

What easy-to-obtain imaging study can cinch the diagnosis?
- b-scan ultrasonography

What will b-scan reveal?
- Choroidal thickening
- Sub-Tenon’s edema
Scleritis

Posterior scleritis: Sub-Tenon’s edema
Scleritis

What easy-to-obtain imaging study can cinch the diagnosis?
- b-scan ultrasonography

What will b-scan reveal?
-- Choroidal thickening
-- Sub-Tenon’s edema

When sub-Tenon’s edema involves the space around the optic nerve, what b-scan finding will result?
Scleritis

Anterior
- Diffuse
- Nodular
  w/ inflammation

Posterior

What easy-to-obtain imaging study can cinch the diagnosis?
b-scan ultrasonography

What will b-scan reveal?
-- Choroidal thickening
-- Sub-Tenon’s edema

When sub-Tenon’s edema involves the space around the optic nerve, what b-scan finding will result?
The classic \textit{T sign}
Scleritis

Posterior scleritis: T-sign
Scleritis

What studies are included in a scleritis work-up?

- ?
- ?
- ?
- ?
- ?
- ?
- ?
- ?
- ?
- ?
What studies are included in a scleritis work-up?
--Serum autoantibodies
--Complete blood count with diff
--Urinalysis
--Syphilis labs
--TB testing
--Serum uric acid
--Sarcoid screening
What studies are included in a scleritis work-up?
--Serum autoantibodies: ? [5 studies]
--Complete blood count with diff
--Urinalysis
--Syphilis labs
--TB testing
--Serum uric acid
--Sarcoid screening

Anterior

Diffuse

Nodular

Scleritis

w/ inflammation

w/o inflammation

Next Q
Scleritis

What studies are included in a scleritis work-up?
--Serum autoantibodies: RF, anti-CCP Ab, ANA, ANCA, anti-DNA
--Complete blood count with diff
--Urinalysis
--Syphilis labs
--TB testing
--Serum uric acid
--Sarcoid screening

w/ inflammation    w/o inflammation
What studies are included in a scleritis work-up?
- Serum autoantibodies: RF, anti-CCP Ab, ANA, ANCA, anti-DNA
- Complete blood count with diff
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RF = ?
CCP
ANA
ANCA
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RF = rheumatoid factor
CCP = ?
ANA
ANCA
What studies are included in a scleritis work-up?

--Serum autoantibodies: RF, anti-CCP Ab, ANA, ANCA, anti-DNA
--Complete blood count with diff
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--TB testing
--Serum uric acid
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RF = rheumatoid factor
CCP = cyclic citrullinated peptide
ANA = ?
ANCA
What studies are included in a scleritis work-up?
--Serum autoantibodies: RF, anti-CCP Ab, ANA, ANCA, anti-DNA
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--Urinalysis
--Syphilis labs
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RF = rheumatoid factor
CCP = cyclic citrullinated peptide
ANA = antinuclear antibody
ANCA = ?
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RF = rheumatoid factor
CCP = cyclic citrullinated peptide
ANA = antinuclear antibody
ANCA = antineutrophil cytoplasmic antibody
What studies are included in a scleritis work-up?
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--Syphilis labs
--TB testing
--Serum uric acid
--Sarcoid screening: ? [5 studies]
Scleritis

What studies are included in a scleritis work-up?
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--Urinalysis
--Syphilis labs
--TB testing
--Serum uric acid
--Sarcoid screening: CXR; ACE, lysozyme, Ca^{2+}, alk phos
Scleritis

What studies are included in a scleritis work-up?
--Serum autoantibodies: RF, anti-CCP Ab, ANA, ANCA, anti-DNA
--Complete blood count with diff
--Urinalysis
--Syphilis labs
--TB testing
--Serum uric acid
--Sarcoid screening: CXR; ACE, lysozyme, Ca²⁺, alk phos

But remember—the most important source of diagnostic clues for any sort of ocular inflammation (scleritis, uveitis, PUK, etc) is a very careful and thorough review of systems.
But remember—the most important source of diagnostic clues for *any* sort of ocular inflammation (scleritis, uveitis, PUK, etc) is a very careful and thorough **review of systems**.
How is scleritis treated?

Systemically

With what?

Diffuse scleritis might respond to PO NSAIDs—try these first. For the others, PO steroids are usually the first-line med. More powerful immunosuppression is frequently required.

What about depot steroids?

Long considered contraindicated, subconjunctival depot steroids have gained wide acceptance as a treatment for scleritis.
Scleritis

How is scleritis treated?
Systemically
How is scleritis treated?
Systemically

With what?
How is scleritis treated?
Systemically

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