Q

CNVM DDx:

- ARMD
- OHS
- Pathologic myopia
- Angioid streaks
- Idiopathic
- Sorsby macular dystrophy (SMD)
- Traumatic choroidal rupture
- Iatrogenic
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ARMD is addressed extensively in a series of slide-sets—see the Table of Contents.
What does OHS stand for in this context?

Ocular histoplasmosis syndrome, aka POHS (the P is for presumed).

Is there a gender predilection in OHS?
No

Is there a racial predilection?
Yes, OHS occurs almost exclusively among whites of Northern European heritage.

Is there a geographic predilection?
Yes, the majority of cases are found in people who reside in the Mississippi/Ohio River valleys of the US.

Does OHS manifest unilaterally, or bilaterally?
Bilaterally (although it can be somewhat asymmetric).

Is OHS associated with vitritis?
Never. If vitritis is present, it's not OHS.

What about AC cell?
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Discrete, focal, atrophic scars.

**Where are they typically located?**
They can be anywhere in the posterior pole, i.e., macula or near-to-mid periphery.

**Are they usually larger, or smaller than the ONH?**
Smaller.

**What two-word phrase is used to describe them?**
‘Punched out’.

**Do they evolve over time?**
Generally no.

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In another nutshell, what is PPA?
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In the last nutshell, what are disciform lesions? --Active lesions represent…either the presence of CNVM under the retina, or a hemorrhagic retinal detachment
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What are you looking for on DFE?
The so-called ‘classic triad’ of OHS

What is the classic triad of OHS?
--Histo spots
--Peripapillary atrophy
--Disciform macular lesion(s)

In the last nutshell, what are disciform lesions?
--Active lesions represent…either the presence of CNVM under the retina, or a hemorrhagic retinal detachment; whereas
--Inactive lesions (aka disciform scars) are…fibrovascular remnants of previous CNVM and/or subretinal hemorrhage

What about vitritis?
Never. If vitritis is present, it’s not OHS.

What about AC cell?
Never. If AC cell is present, it’s not OHS.
CNVM DDx

Active lesion

Disciform scar

OHS
What does OHS stand for in this context?
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--Histo spots?
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--Disciform macular lesion(s)?

What is the classic triad of OHS?
In the last nutshell, active lesions represent either the presence of CNVM under the retina, or a hemorrhagic retinal detachment. Whereas inactive lesions, also known as disciform scars, are fibrovascular remnants of previous CNVM and/or subretinal hemorrhage.

Which lesion(s) require treatment?
Only active disciform lesions.

What treatment modalities are used to treat active disciform lesions?
--Thermal laser
--Photodynamic therapy (PDT)
--Anti-VEGF therapy
--Submacular surgery
--Intravitreal corticosteroids
--Combination therapy (of some of the above modalities)

Can OHS manifest unilaterally, or bilaterally?
Bilaterally (although it can be somewhat asymmetric).

Is OHS associated with vitritis?
Never. If vitritis is present, it’s not OHS.

What about AC cell?
Never. If AC cell is present, it’s not OHS.

Is there a gender predilection in OHS?
No.

Is there a racial predilection?
Yes, OHS occurs almost exclusively among whites of Northern European heritage.

Is there a geographic predilection?
Yes, the majority of cases are found in people who reside in the Mississippi/Ohio River valleys of the US.
**CNVM DDx**

- ARMD
- **OHS**
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy (SMD)
- Traumatic choroidal rupture
- Iatrogenic

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**Which lesion(s) require treatment?**
Only active disciform lesions.

**In the last nutshell**, we can probably dispel the notion of a homogenous retinal involvement:
--**Inactive lesions** (aka previous CNVM and/or subretinal hemorrhage),
--**Active lesions** (either the presence of CNVM under the retina, or a hemorrhagic retinal detachment).
**CNVM DDx**

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**In the last nutshell, what are the treatment modalities?**
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The so-called ‘classic triad’ of OHS:

- Histo spots
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What is the classic triad of OHS?

In the last nutshell, all the following:

- Active lesions represent either the presence of CNVM under the retina, or a hemorrhagic retinal detachment.
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Which lesion(s) require treatment?
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**Do antifungals play a role in the treatment of OHS?**

--Antifungals?
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--Histio spots
--Peripapillary atrophy
--Disciform macular lesion(s)

In the last nutshell, we have the so-called ‘classic triad’ of OHS: histio spots, peripapillary atrophy, and disciform macular lesion(s). The disciform lesion(s) is/are the hallmark of OHS.

Which lesion(s) require treatment?
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What treatment modalities are used to treat active disciform lesions?
--Thermal laser
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- **Antifungals? No!**

**Do antifungals play a role in the treatment of OHS?**
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What is the classic DFE appearance of angioid streaks?

Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch's membrane.
Q/A

- **CNVM DDx:**
  - ARMD
  - POHS
  - **Angioid streaks**

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What proportion of angioid streaks are associated with systemic abnormalities? About half.

What is the well-known mnemonic for angioid streak's associations? What are these associations?

- **P**seudoxanthoma elasticum (PXE)
- **E**hlers-Danlos syndrome
- **P**aget's disease of bone
- **S**ickle-cell disease
- **I**diopathic
What is the classic DFE appearance of angioid streaks?
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Angioid streaks (arrowheads). Note that only a few of the many present have been marked.
• **CNVM DDx:**
  - ARMD
  - POHS
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- **P**aget’s disease of bone
- **S**ickle-cell disease
- **I**diopathic (ie, no association)

~50% of cases are associated with one of these.

~50% of cases have no known systemic association.

*(No question, proceed when ready)*
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Idiopathic (ie, no association)

Which condition has the strongest association with angioid streaks?
CNVM DDx:

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Which condition has the strongest association with angioid streaks?
PXE, by a mile.

Briefly, what sort of disorder is PXE?

An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues.

Is it common, or rare?

Rare.

Is there a gender predilection?

Yes, ♀ are twice as likely to be affected.

Other than the eye, what organ-systems are affected?

--Skin
--Vascular system
--GI tract
--Eye.
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Skin, vascular system, GI tract, eye.
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**Is it common, or rare?**
Rare

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Yes, ♀ are twice as likely to be affected.

**What is the appearance of affected skin?**
An area of waxy-yellow, papule-like lesions.

**What is the classic informal descriptor for this appearance?**
‘Chicken skin’

**In what two locations is ‘chicken skin’ most often found?**
--The neck
--The axillae

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PXE skin

Angioid Streaks
Q

CNVM DDx

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Other than the eye, what organ-systems are affected?
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There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?
--Angioid streaks
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**CNVM DDx**

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- RPE mottling
- Optic disc drusen

**Briefly, what sort of disorder is PXE?**
An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues.

**Is it common, or rare?**
Rare.

**Is there a gender predilection?**
Yes, ♀ are twice as likely to be affected.

**About the eye, what organ-systems are affected?**
- Skin
- Vascular system
- GI tract
- Eye

**What is the well-known mnemonic for angioid streak’s associations? What are these associations?**
- Pseudoxanthoma elasticum (PXE)
- Ehlers-Danlos syndrome
- Paget’s disease of bone
- Sickle-cell disease
- Idiopathic (ie, no association)
**Q**

**CNVM DDx**

- **Pseudoxanthoma elasticum (PXE)**
- Ehlers-Danlos syndrome
- Paget’s disease of bone
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- Idiopathic (ie, no association)

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**Is there a gender predilection?**
Yes, ♀ are twice as likely to be affected.

**Other than the eye, what organ-systems are affected?**
- Skin
- Vascular system
- GI tract
- Eye

There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?
- Angioid streaks
- Optic disc drusen

**What mellifluous name is used to describe the RPE mottling?**
Peau d'orange

Which condition has the strongest association with angioid streaks? PXE, by a mile.
CNVM DDx

Briefly, what sort of disorder is PXE?
An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues

Is it common, or rare?
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Which condition has the strongest association with angioid streaks?
PXE, by a mile
PXE: *Peau d’orange* fundus
Briefly, what sort of disorder is PXE?
An elastorrhexis, ie, a condition characterized by progressive calcification and fragmentation of elastic tissues

Is it common, or rare?
Rare

Is there a gender predilection?
Yes, ♀ are twice as likely to be affected

What is the classic DFE appearance of angioid streaks?
Reddish-brown lines radiating out from the peripapillary region; these lines represent breaks in Bruch's membrane

What proportion of angioid streaks are associated with systemic abnormalities?
About half

What is the well-known mnemonic for angioid streak's associations? What are these associations?
Pseudoxanthoma elasticum (PXE)
Ehlers-Danlos syndrome
Paget’s disease of bone
Sickle-cell disease
Idiopathic (ie, no association)

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There are three classic eye findings in PXE, one of which is angioid streaks. What are the other two?
Angioid streaks
RPE motting
Optic disc drusen

For more on angioid streaks, see slide-set R61
Q

CNVM DDx:

- ARMD
- POHS
- Angioid streaks
- **Pathologic myopia**
- Idiopathic
- Sorsby macular dystrophy
- Choroidal rupture after trauma
- Iatrogenic

*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?*
• CNVM DDx:
  • ARMD
  • POHS
  • Angioid streaks
  • **Pathologic myopia**
  • Idiopathic
  • Sorsby macular dystrophy
  • Choroidal rupture after trauma
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- Sorsby macular dystrophy
- Choroidal rupture after trauma
- Iatrogenic

What proportion of eyes longer than 26.5 mm will develop CNV?

About 10%

What axial length serves as a useful cutoff for defining pathologic myopia?

26.5 mm
A

- CNVM DDx:
  - ARMD
  - POHS
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  - Pathologic myopia
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?*

- 26.5 mm

*What is the classic finding on DFE that puts high myopes at risk for CNVM?*
CNVM DDx:

- ARMD
- POHS
- Angioid streaks
- **Pathologic myopia**
- Idiopathic
- Sorsby macular dystrophy
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- Iatrogenic

**Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?**
26.5 mm

**What is the classic finding on DFE that puts high myopes at risk for CNVM?**
Lacquer cracks
Q

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia

Angioid streaks vs lacquer cracks: Compare and contrast

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26.5 mm

What is the classic finding on DFE that puts high myopes at risk for CNVM?
Lacquer cracks
Q/A

- CNVM DDx:
  - ARMD
  - POHS
  - **Angioid streaks**
  - Pathologic myopia

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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
Angioid streaks
‘Reddish-brown’

Lacquer cracks
‘Yellowish’
Q/A

- **CNVM DDx:**
  - ARMD
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Q/A

- **CNVM DDx:**
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*Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm*
Angioid streaks
‘Reddish-brown’
‘Peripapillary’

Lacquer cracks
‘Yellowish’
‘Macular’
Q&A

CNVM DDx:
- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Sorsby macular dystrophy (SMD)
- Choroidal rupture after trauma
- Iatrogenic

Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm

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- CNVM DDx:
  - ARMD
  - POHS
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  - Pathologic myopia

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Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm

What is the classic finding on DFE that puts high myopes at risk for CNVM? Angioid streaks
Q/A

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia

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Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia?
26.5 mm

What is the classic finding on DFE that puts high myopes at risk for CNVM?
Lacquer cracks
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Per the Retina book, what axial length serves as a useful cutoff for defining pathologic myopia? 26.5 mm.
Q

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia
  - Idiopathic
  - **Sorsby macular dystrophy**
  - Choroidal rupture
  - Iatrogenic

*In two words, what sort of condition is Sorsby?*  
*(Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)*
CNVM DDx:

- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
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In two words, what sort of condition is Sorsby? (Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)

A macular dystrophy
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In two words, what sort of condition is Sorsby? (Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)

A **macular dystrophy**

For context and completeness’ sake: What are the other macular dystrophies with which Sorsby’s is addressed?

- -
- -
- -
- -
- -
- -

**Bilateral subfoveal CNVMs at age 40**
A

- CNVM DDx:
  - ARMD
  - POHS
  - Angioid streaks
  - Pathologic myopia
  - Idiopathic
  - Sorsby macular dystrophy
  - Choroidal rupture
  - Iatrogenic

In two words, what sort of condition is Sorsby?
(Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)

A macular dystrophy

For context and completeness’ sake: What are the other macular dystrophies with which Sorsby’s is addressed?
--Stargardt
--Best vitelliform
--Adult-onset vitelliform
--The ‘pattern’ dystrophies
--Central areolar choroidal dystrophy
--North Carolina macular dystrophy
CNVM DDx:

- ARMD
- POHS
- Angioid streaks
- Pathologic myopia
- Idiopathic
- **Sorsby macular dystrophy**
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- Iatrogenic

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A macular dystrophy

*What is the inheritance pattern?*
A

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**CNVM DDx**

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*What is the inheritance pattern?*

AD
CNVM DDx:
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**Q**

In two words, what sort of condition is Sorsby? *(Hint: It’s the two-word header of the section in the Retina book in which Sorsby is discussed.)*

A macular dystrophy

What is the inheritance pattern?
AD

What is the classic DFE finding in a pt who has the condition, but has yet to become symptomatic? *(Hint: It’s indicated by the subheader of the section in the Retina book in which it is presented.)*

A young adult with large numbers of “drusenlike deposits” in the maculae

What is the classic presentation of a pt who has become symptomatic?
Bilateral subfoveal CNVMs at age 40
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Sorsby macular dystrophy
Sorsby: Drusenlike deposits
Q

- CNVM DDx:
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Q/A

- CNVM DDx:
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**CNVM DDx**

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*What is the classic presentation of a pt who has become symptomatic?*

Bilateral subfoveal CNVMs at age 40

# laterality, and location  CNVMs at age
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Bilateral subfoveal CNVMs at age 40

The story you’re looking for (on a test) is of an adult who had good vision bilaterally until their early 40s, when they noted sudden VA decrease in one eye, then the other. DFE and imaging will reveal CNVM in the affected eye(s) if vision loss is recent, extensive scarring if remote.

*(No question, proceed when ready)*
Sorsby: Late extensive fibrosis/scarring
Q

- CNVM DDx:
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What is the subheader name?
‘Early-onset “drusenoid” macular dystrophies’

What is the classic presentation of a pt who has become symptomatic?
Bilateral subfoveal CNVMs at age 40