In a nutshell, what is scleritis?
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?
Scleritis

In a nutshell, what is scleritis?
An inflammatory condition characterized by painful infiltrative scleral edema

Is it serious, or more of a nuisance?
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
Scleritis

Very basic anatomic division
Scleritis

Very basic anatomic division

Anterior

Posterior
Scleritis

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
Scleritis

Scleritis demographics:
- Children < Adults
- Men < Women

Unilateral ≈ Bilateral

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

Posterior
Scleritis

Scleritis demographics:
Children << Adults

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(Not necessarily simultaneously; just at some point during the disease course)
Scleritis

Scleritis demographics:
- Children << Adults
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- Unilateral ≈ Bilateral

(Not necessarily simultaneously; just at some point during the disease course)
What are three classic signs of anterior scleritis?

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

- Anterior
  - Diffuse
  - Nodular
  - Necrotizing

- Posterior

Is the nodule of nodular scleritis mobile?

w/o inflammation
Scleritis

Anterior

- Diffuse
- **Nodular**

Posterior

- Necrotizing

*Is the nodule of nodular scleritis mobile?*

**No**
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?
If a patient has a necrotizing scleritis with inflammation, it’s a virtual lock that what sort of disease is the culprit? A vasculitis
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?
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?*

*No, but it certainly can be*
If a patient has a necrotizing scleritis w/ inflammation, it's a virtual lock that what sort of disease is the culprit?

A **vasculitis**

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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?

No, but it certainly can be

Why is this so important?

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. The presence of ocular vasculitis in systemic disease portends a dismal prognosis for the patient. Five-year mortality for such patients approaches 50%!
Scleritis

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?

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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.

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Scleritis

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

- **Posterior**
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.
By what other name is necrotizing scleritis w/o inflammation known?
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Scleritis

Anterior

Diffuse Nodular Necrotizing

w/ inflammation w/o inflammation
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Does scleromalacia perforans occur early in the RA dz course?
- No, it tends to occur in longstanding cases
Scleritis

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

Anterior

Diffuse
Nodular

From what related disease must scleritis be differentiated?

Scleritis

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

Scleritis

Episcleritis

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

Episcleritis

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

½ w/ systemic condition
½ no systemic condition

(2 most common classes?)
What proportion of scleritis patients have an identifiable systemic condition? About 1/2
What proportion of scleritis patients have an identifiable systemic condition? About 1/2
Scleritis

Anterior
Diffuse  Nodular  Necrotizing
w/ inflammation  w/o inflammation

Posterior

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

½ w/ systemic condition  ½ no systemic condition

CT Disease  Vasculitis
RA (rheumatoid arthritis)  SLE (systemic lupus erythematosus)  SNSAs (seronegative spondyloarthropathies)
What proportion of scleritis patients have an identifiable systemic condition? About 1/2

To what does the term seronegative spondyloarthropathies refer?

- Ankylosing spondylitis
- Reactive arthritis
- Psoriatic arthritis
Scleritis

Anterior

Posterior

Diffuse

Nodular

Necrotizing

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

½ no systemic condition
½ with systemic condition

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- RA (rheumatoid arthritis)
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To what does the term seronegative spondyloarthropathies refer?
It is an umbrella term for a group of related rheumatologic conditions

To what does the term seronegative refer?
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

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

½ no systemic condition
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Vasculitis

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About 1/2 with systemic condition

½ no systemic condition

Scleritis

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Scleritis

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½ no systemic condition
½ w/ systemic condition

CT Disease
- RA
- SLE
- SNSAs

Vasculitis

(3 most common vasculitides?)
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

½ no systemic condition
½ with systemic condition

CT Disease
- RA
- SLE
- SNSAs

Vasculitis
- Wegener’s
- PAN
- GCA

Which of these diseases are associated with episcleritis?

Episcleritis

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 have no systemic condition, and ½ with a systemic condition.

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.
Scleritis

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.

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.
What proportion of scleritis patients have an identifiable systemic condition?

About 1/2

Anterior Posterior

Diffuse Nodular

w/o inflammation

w/ inflammation

Scleritis

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Why surgical debridement? That is, why aren’t abx enough?
**Scleritis**

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Anterior Posterior

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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**

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

--

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 uveitis to be associated with a systemic condition?

Less likely
Scleritis

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

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

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

**Posterior**

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Less likely
Scleritis

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

- 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 ocular rotations
-- The pain awakens the pt at night
Scleritis

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

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

--

--

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 [two words]

Anterior

Posterior

w/ inflammation  w/o inflammation
Scleritis

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
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--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

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 T sign
Scleritis

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

Posterior

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

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
  - w/ inflammation

Posterior

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- b-scan ultrasonography

What will b-scan reveal?
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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
Scleritis

Anterior

Diffuse

Nodular

Posterior

w/ inflammation

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When sub-Tenon’s edema involves the space around the optic nerve, what b-scan finding will result?
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When sub-Tenon’s edema involves the space around the optic nerve, what *b*-scan finding will result?
The classic *T sign*
What studies are included in a scleritis work-up?

- Serum autoantibodies (RF, ANA, ANCA, Anti-DNA)
- CBC with diff
- UA
- Syphilis studies
- PPD
- Serum uric acid
- Sarcoid screening (CXR, ACE, lysozyme)
Scleritis

What studies are included in a scleritis work-up?
--Serum autoantibodies (RF, ANA, ANCA, Anti-DNA)
--Complete blood count with differential
--Urinalysis
--Syphilis studies
--PPD (i.e., tuberculin skin test)
--Serum uric acid
--Sarcoid screening (chest X-ray, ACE, lysozyme)

ACE = angiotensin converting enzyme
RF = rheumatoid factor
ANA = antinuclear antibody
ANCA = antineutrophil cytoplasmic antibody
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But remember—the most important source of diagnostic clues for any sort of ocular inflammation (scleritis, uveitis, PUK, etc) is a very careful and very 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.

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

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- Nodular
- Necrotizing
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Posterior

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