## Q

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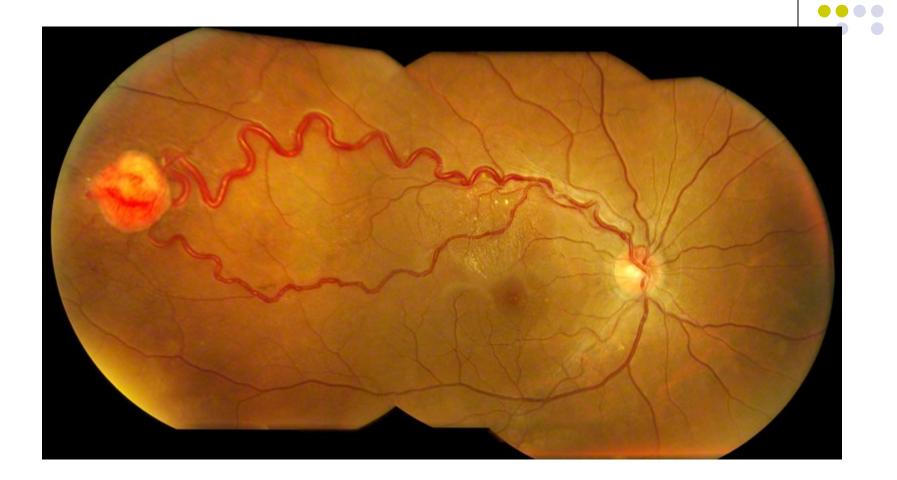
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#### Capillary hemangioblastoma in von Hippel-Lindau dz

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The oddball here is *capillary hemangioblastoma*, which is a retinal lesion associated with **von Hippel-Lindau disease**.

In a word, what sort of condition is von Hippel-Landau disease?



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*In a word, what sort of condition is von Hippel-Landau disease?* A phakomatosis



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Phakomatoses are known also as what sort of syndrome?

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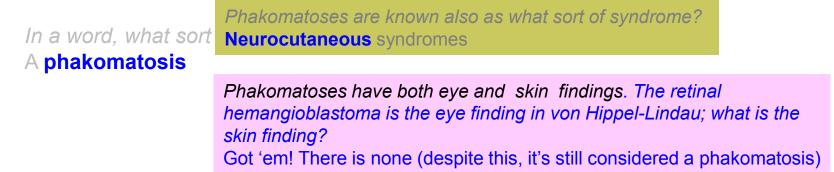
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In general, how will an intraconal mass present?







*In general, how will an intraconal mass present?* With one or (usually) more of the following:

- --?
- . --?

#### Schwannoma



*In general, how will an intraconal mass present?* With one or (usually) more of the following:

- --Proptosis
- --Pain
- --Diplopia

### Schwannoma







Intraconal mass OS producing proptosis



In general, how will an intraconal mass present? With one or (usually) more of the following: --Proptosis --Pain --Diplopia (If the two words is involved, vision will be affected as well)

• Schwannoma





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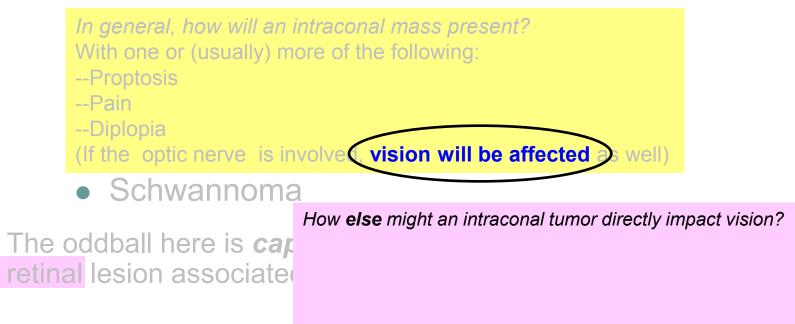
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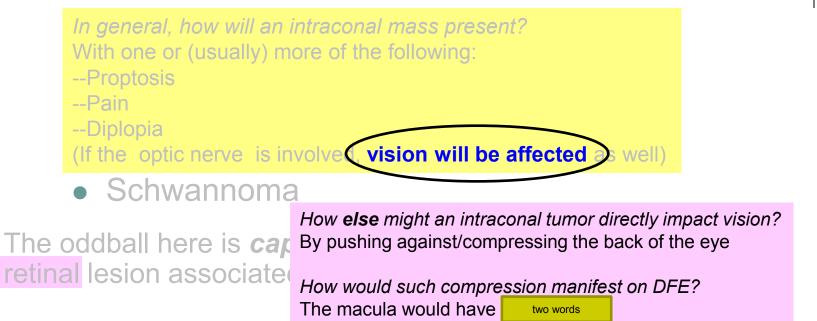






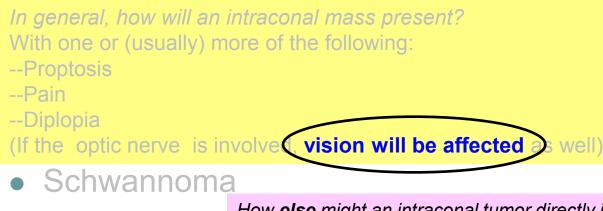












The oddball here is **car** retinal lesion associate

How **else** might an intraconal tumor directly impact vision? By pushing against/compressing the back of the eye

How would such compression manifest on DFE? The macula would have retinal striae







Fundus photo demonstrating retinal striae in a pt with an intraconal cavernous hemangioma compressing the posterior pole





In general, how will an intraconal mass present? With one or (usually) more of the following:

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Schwannoma

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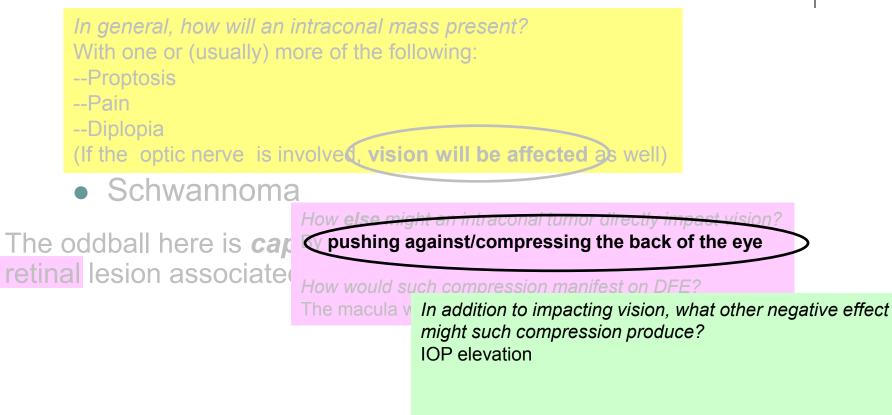
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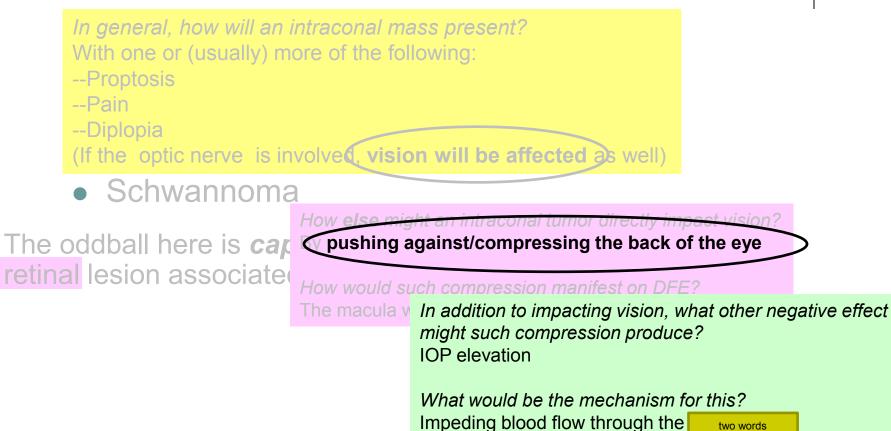
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What would be the mechanism for this?











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> What would be the mechanism for this? Impeding blood flow through the vortex veins



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The oddball here is *capillary hemangioblastoma*, which is a retinal lesion associated with von Hippel-Lindau disease. *Re the others:* 

32

-- intraconal differential tumor is the most common intraorbital tumor in adults, and is usually intraconal.



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FYI: Cavernous hemangiomas are known also by what other name?

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\*One section of the most recent (at the time of this writing) version of the *Plastics* book indicates that *cavernous venous malformations* is now the **preferred** nomenclature. That said, other current *BCSC* volumes continue to use the term cavernous hemangioma (as do other sections of the *Plastics* book, for that matter). Caveat emptor.



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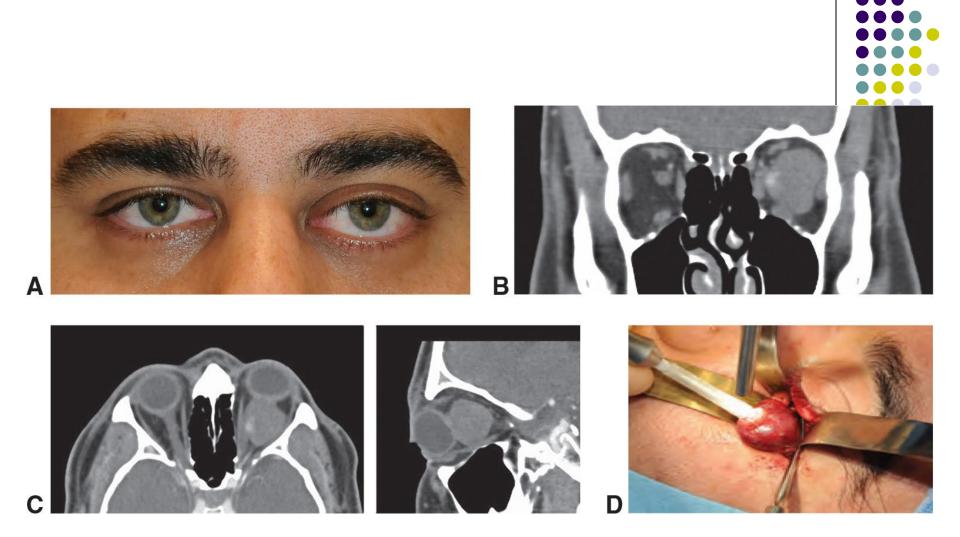
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How does it appear on imaging? As a well-encapsulated mass





47

Cavernous hemangioma. *A*, Proptosis of the left eye as a result. *B*, Coronal CT shows a well-circumscribed lesion the muscle cone. *C*, Axial *(left)* and sagittal *(right)* CT show the mass. *D*, Intraoperative traction with a cryoprobe facilitates complete removal of the mass.

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For more on cavernous hemangiomas, see slide-set O10

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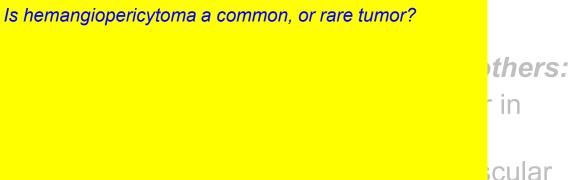
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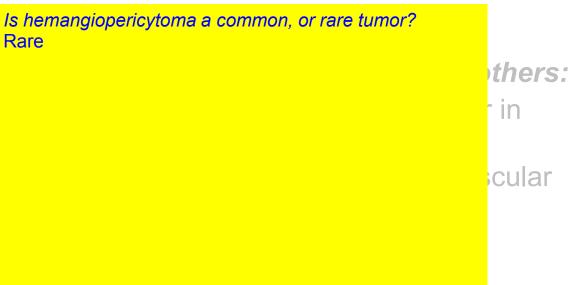
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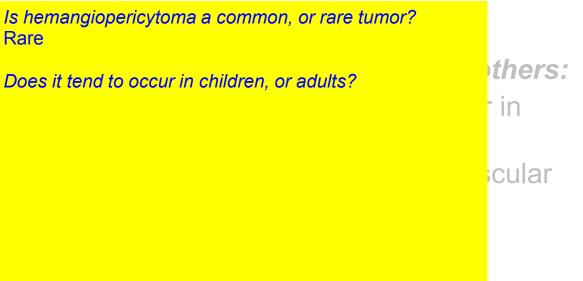
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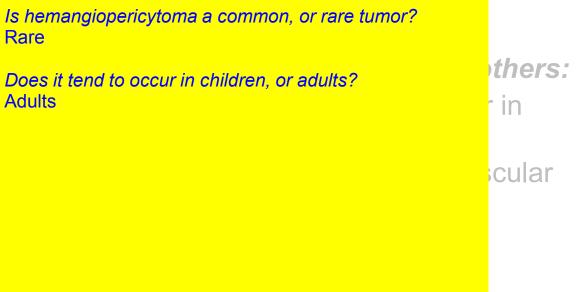
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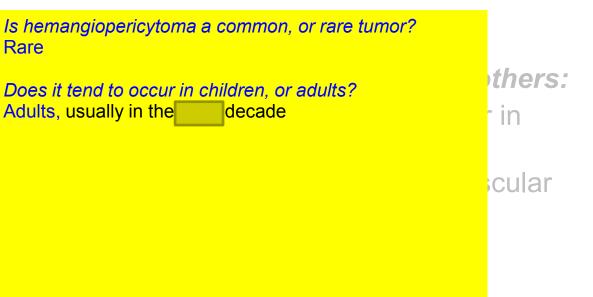
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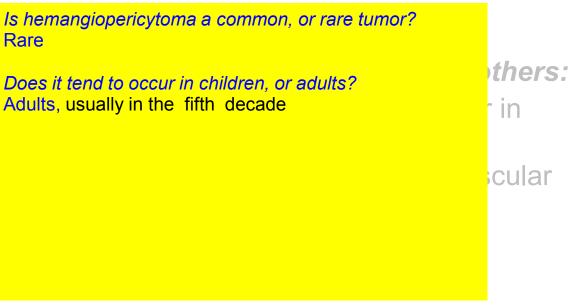
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*Is hemangiopericytoma a common, or rare tumor?* Rare *Does it tend to occur in children, or adults?* 

Adults, usually in the fifth decade

How does it appear on imaging?



*thers:* in cular

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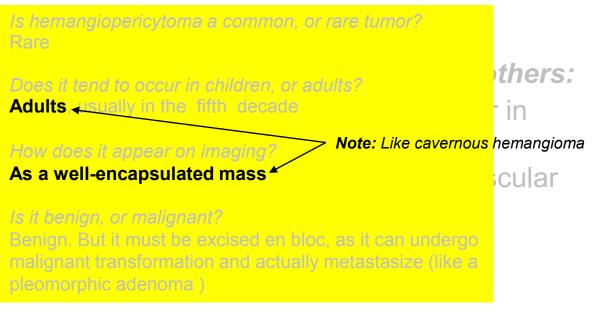
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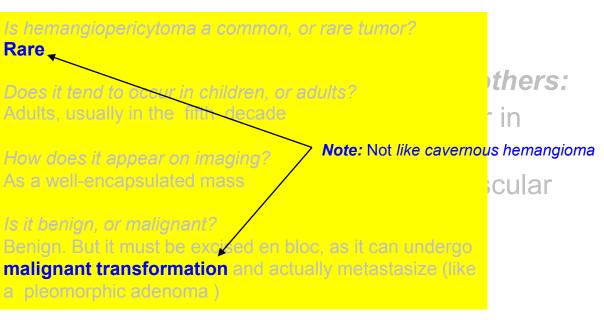
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---Cavernous hemangioma is the most common intraorbital tumor in adults, and is usually intraconal.

--Hemangiopericytoma is a locally aggressive proliferation of vascular pericytes.

intraconal differential tumor

is a mesenchymal tumor that can be benign, locally aggressive or metastatic.

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*Is fibrous histiocytoma a common, or uncommon entity?* Uncommon

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The oddball here is *capillary hemangioblastoma*, which is a retinal lesion associated with von Hippel-Lindau disease. *Re the others:* 

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