

Cavernous Hemangioma: Painless Proptosis in a 62 year-old Woman

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Chief Complaint: "My right pupil is higher than my left pupil"

History of Present Illness:

A 62 year-old woman with a one-year history of breast carcinoma status post lumpectomy, chemotherapy and radiation presents to the general ophthalmology clinic for routine follow-up of a symptomatic posterior vitreous detachment in her left eye. She states that over the last year she has noticed that her right pupil is higher than her left. Additionally, she wonders why more sclera is showing under her right iris than under her left. She denies pain or double vision.

Past Ocular History:

- Dry eye
- Ocular rosacea
- Refractive error/presbyopia
- Mild cataracts

Past Medical History:

Infiltrating ductal carcinoma of the breast (Elston-Ellis grade III, ER/PR +, HER-2/neu +) status post right breast lumpectomy with lymph node dissection, chemotherapy and radiation

Medications: Tetracycline, vitamins

Allergies: Morphine, Keflex (cephalexin), codeine, Talwin (pentazocine lactate), Taxol (paclitaxel), mold, milk

Family History: Thyroid disease (mother and sister), post-menopausal breast cancer (mother and maternal aunt), lung cancer (father), prostate cancer (brother), ovarian cancer (sister)

Social History: Works as a nursing coordinator at a local college

Physical Exam:

- Visual acuity:
 - 20/20 -1 OD
 - 20/20 -2 OS

- Current spectacle correction:
 - OD: + 0.25 + 0.50 x 060
 - OS: plano + 0.50 x 001
- Pupils: 4 mm → 2 mm, briskly reactive without RAPD OU
- Extraocular motility: Full OU
- Confrontational visual fields: No deficits OU
- Intraocular pressure:
 - 19 mmHg OD
 - 21 mmHg OS
- External examination: Right hyperglobus with inferior scleral show
- Anterior segment examination: Meibomian gland dysfunction OU, trace punctate epithelial erosions OS, mild nuclear sclerosis OU, otherwise normal examination
- Fundus examination: Normal OU

Clinical course: The patient was referred to the oculoplastics service and found to have 2 mm of right eye proptosis as well as right hyperglobus with inferior scleral show. Given her history of cancer, there was concern for a possible metastatic orbital mass. Contrast-enhanced CT of the orbits was performed and showed a well-demarcated 1.6 cm soft tissue density in the inferior lateral aspect of the right orbit. There was some minimal scalloping of adjacent lateral orbital wall. Although the mass had a benign appearance, the patient requested removal after reviewing the risks and benefits.



Figure 1: 62 year-old woman with painless proptosis noticeable over the last year. Right hyperglobus leading to inferior scleral show is evident.

Maxillofacial CT with contrast

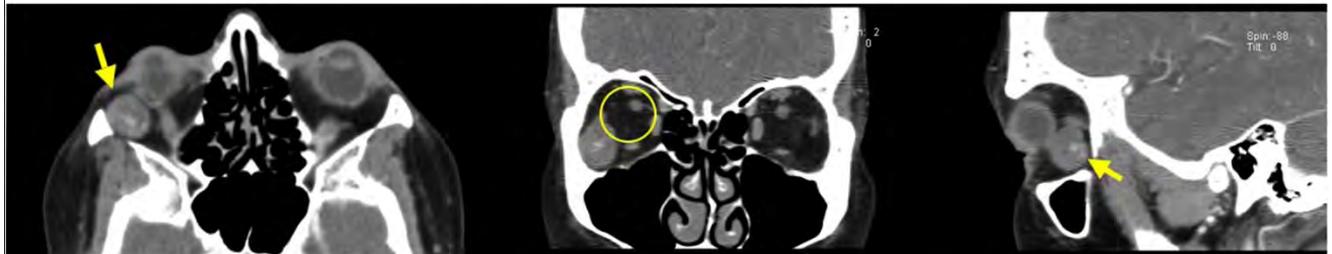


Figure 2: Maxillofacial CT demonstrates a 1.6 cm somewhat heterogenous extraconal round mass inferolaterally in the right orbit (arrows). The yellow circle delineates the margins of the muscle cone.

A right lateral orbitotomy was performed with excisional biopsy of the mass. Pathologic examination showed an encapsulated mass consisting of irregular, thin-walled vascular spaces. The spaces were filled with red blood cells. Radiographic and pathologic findings were consistent with cavernous hemangioma of the orbit.

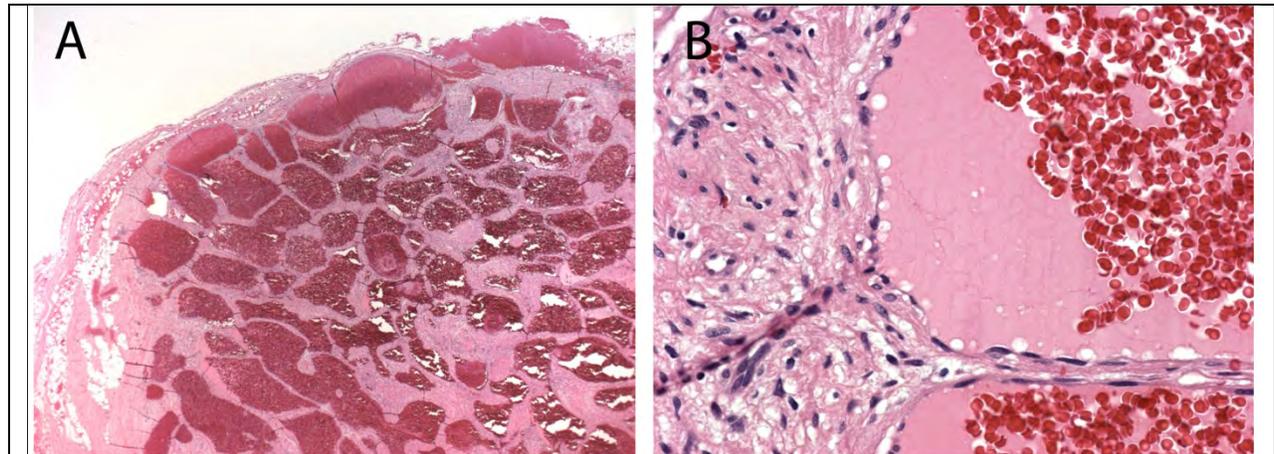


Figure 3: **A**, 10x H&E stained section showing an encapsulated mass with multiple cavernous spaces separated by fibrous septae. **B**, 200x section showing RBCs in the cavernous space, which is lined by mature endothelial cells. (photos compliments of Nasreen Syed, MD, The F.C. Blodi Eye Pathology Laboratory, The University of Iowa)

Discussion:

Cavernous hemangioma is the most common primary orbital tumor in adults (Cohen 2009). It is classically a well-circumscribed unilateral intraconal mass seen in middle-aged women. It is not uncommon for these to be found incidentally after imaging is performed for another condition (e.g. headaches, seizures, etc.). The clinical hallmark is a slowly progressive, painless proptosis, but patients may also present with vision loss either due to optic nerve compression or hyperopic shift when the mass compresses the posterior pole. In the latter case, choroidal folds may be seen on fundus exam. As with any space occupying lesion in the orbit, patients may also note diplopia. Given the slow progression of cavernous hemangiomas, patients, as ours did, may be able to fuse deviations that would have produced diplopia if onset were acute.

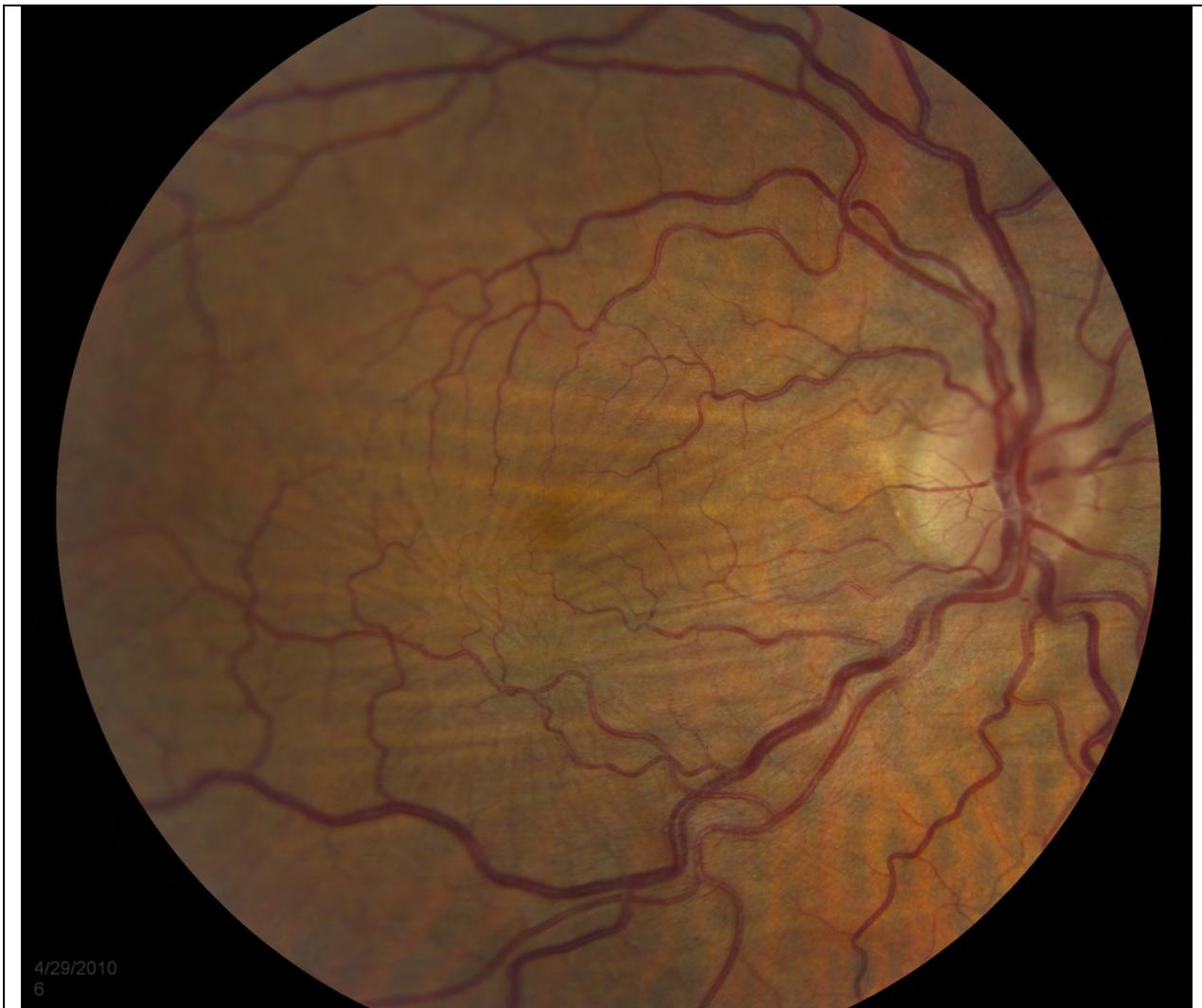


Figure 4: Color fundus photograph of the right eye demonstrating choroidal folds in a patient with an intraconal cavernous hemangioma compressing the posterior pole.

Histopathologically, these tumors are thought to be congenital vascular hamartomas, though there is some speculation regarding an acquired etiology. They account for approximately 80% of orbital vascular tumors and invariably take a benign course in contrast to hemangiopericytomas, which can mimic cavernous hemangioma both in presentation and on imaging (Kikuchi et al 1994).

CT, MRI and echography have all been studied for the evaluation of cavernous hemangioma (Thorn-Kany et al 1999, Dallaudiere et al 2009). Choice of imaging depends on clinical suspicion, but the authors feel that CT with contrast is a good first choice as this will yield information about the shape, vascularity, consistency, and margins of the lesion as well as adjacent bony changes. Some studies report that MRI can be helpful in differentiating between schwannoma, hemangiopericytoma, and

cavernous hemangioma (Xian et al 2010, Kikuchi 1994). Coronal images are useful to assess position of the lesion relative to the optic nerve (see Figure 9). Some general imaging characteristics are highlighted below.

CT appearance: Well-defined oval or round mass roughly isodense to muscle. Hyperdense areas within the lesion may correspond to areas of thrombosis termed "phleboliths." Contrast enhancement is homogenous, but fills slowly.

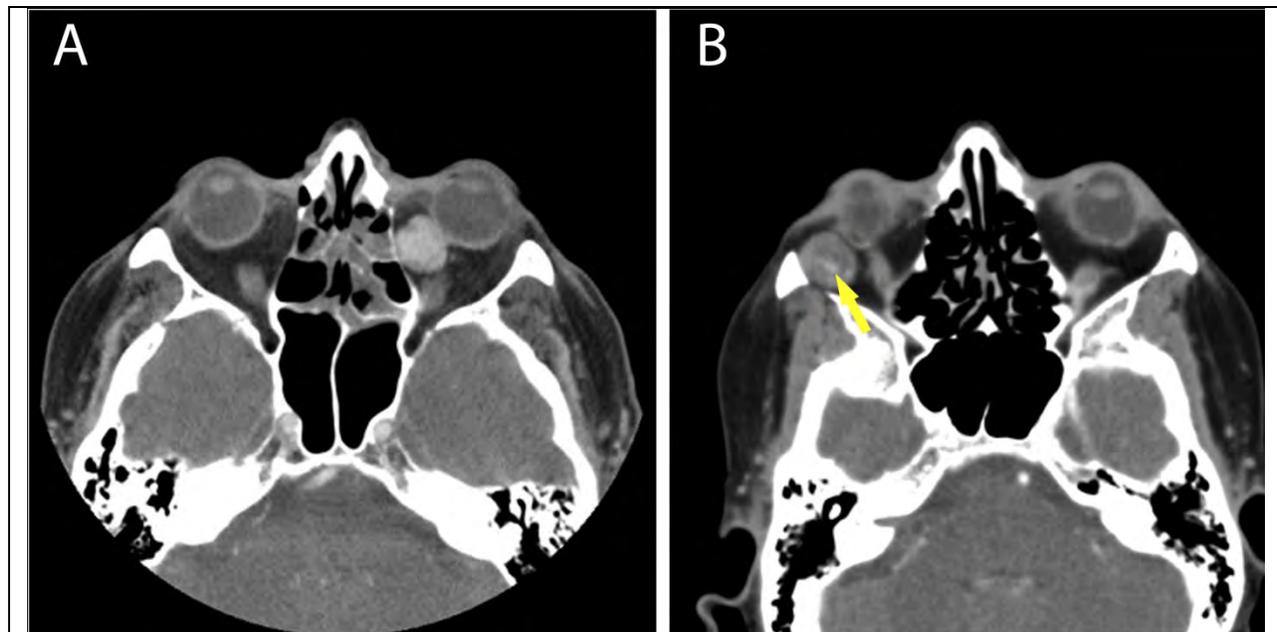


Figure 5: A, Axial CT with contrast showing a well demarcated round lesion in the medial orbit. Analysis of other cuts demonstrates this to be an intraconal lesion. B, Axial CT with contrast in our patient with possible phlebolith (arrow).

MRI appearance: T1-weighted imaging: Homogeneous signal isointense to muscle.

T1 Weighted Imaging (fat suppressed)

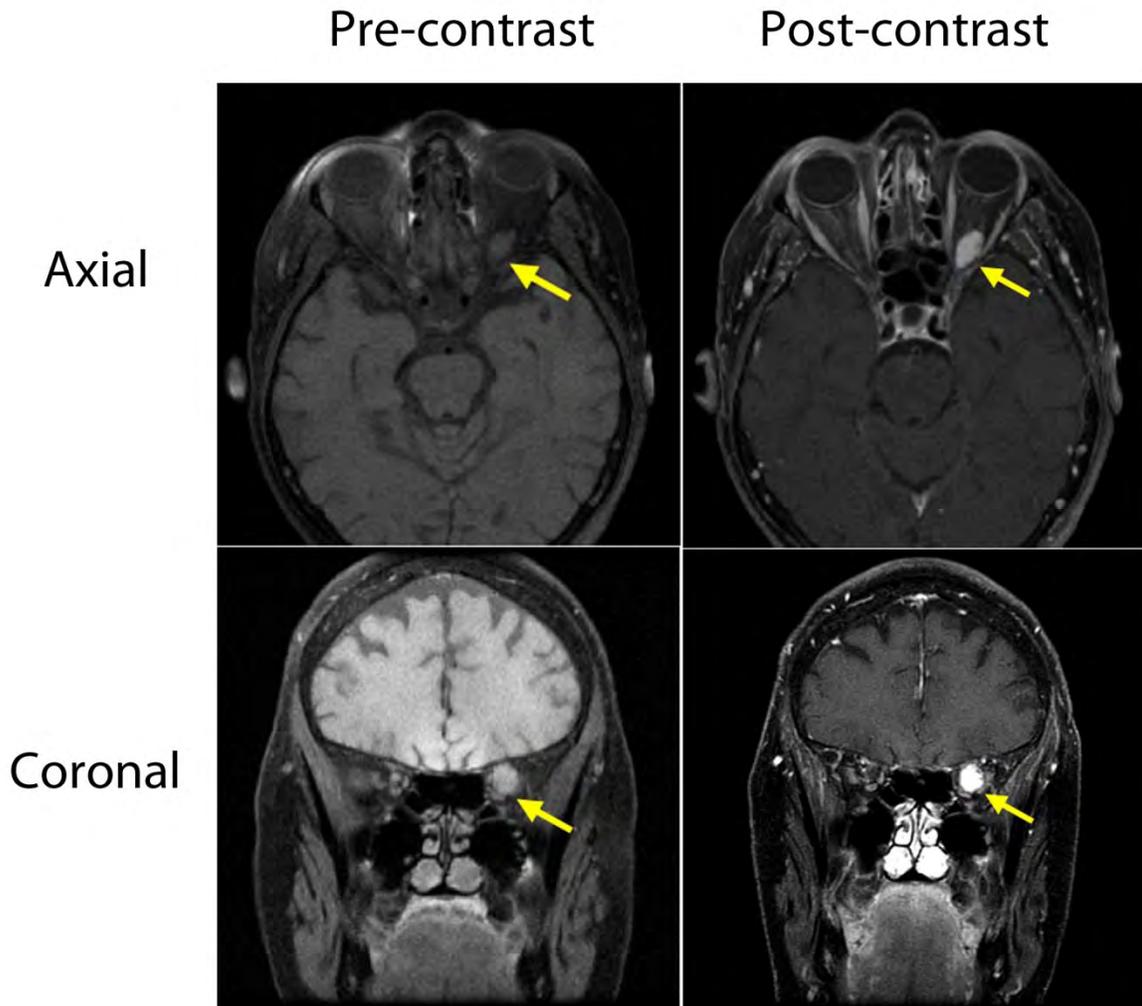


Figure 6: T1 pre and post contrast images with fat suppression. Gadolinium is helpful to assess the vascularity of the lesion. Post-gadolinium enhancement is usually homogenous in cavernous hemangiomas as seen here, though often delayed.

T1 Weighted Image (without fat suppression)



Figure 7: T1 image without fat suppression or contrast. Without gadolinium, the hemangioma is best seen without fat suppression on T1 as hyperintense fat helps demarcate the less intense lesion.

T2 weighted imaging: Homogeneous signal hyperintense to muscle best seen with fat suppression.

T2 Weighted Image (fat suppressed)

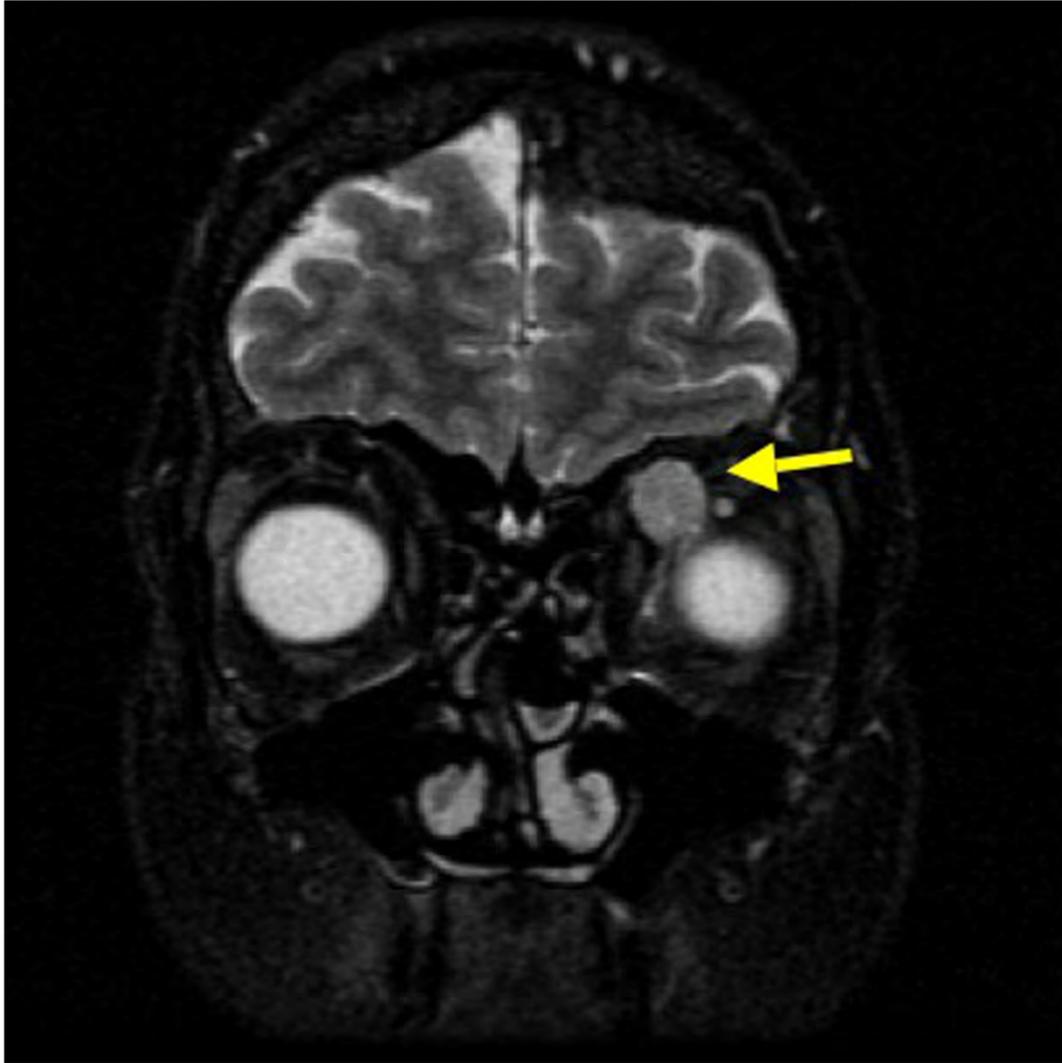


Figure 8: T2 fat suppressed image showing a well demarcated hyperintense mass in the superomedial orbit.

In summary, lesions are easiest to see either on non-contrast-enhanced T1 images without fat suppression, fat-suppressed T2 images, or T1 fat-suppressed contrast-enhanced images (Ruchman and Flanagan 1983, Thorn-Kany et al 1999).

If history, exam and imaging studies are consistent with cavernous hemangioma it is reasonable to observe without obtaining a tissue diagnosis. In the majority of cases there is little change in the appearance of the lesion as demonstrated by the CT scan below. However, patients should be monitored with imaging every 1-2 years for the first

five years after diagnosis to confirm stability. If the lesion is stable after five years, then there is no need to reimaging unless there are any new clinical findings.

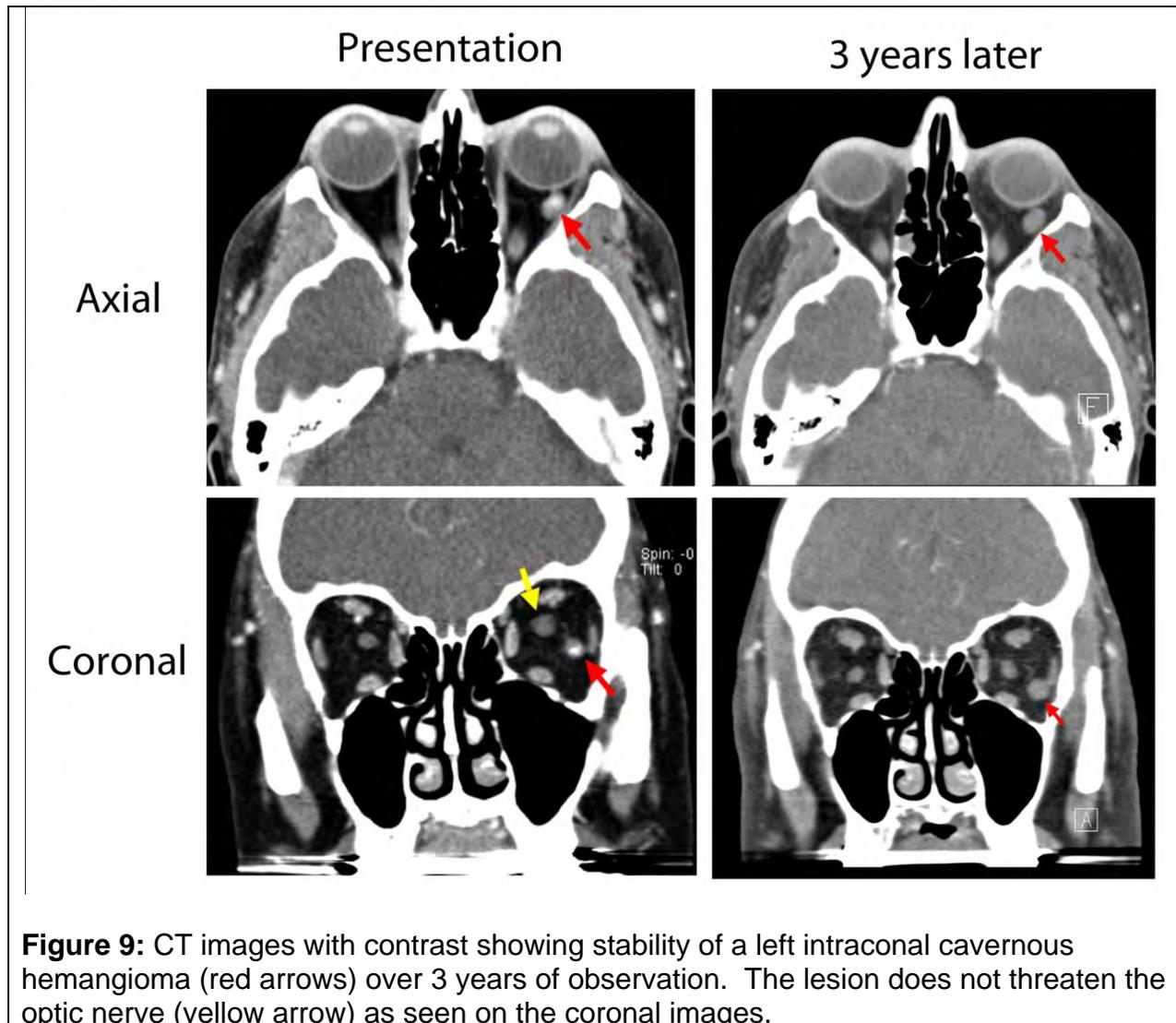


Figure 9: CT images with contrast showing stability of a left intraconal cavernous hemangioma (red arrows) over 3 years of observation. The lesion does not threaten the optic nerve (yellow arrow) as seen on the coronal images.

However, in the setting of known malignancy (such as in this case) or rapid progression in size, biopsy is reasonable to rule out other etiologies for the mass. In addition, diplopia, optic nerve compression and choroidal folds are indications for removal. Excision is usually not necessary depending on the size and location of the lesion and in the absence of vision loss. Complete excision can usually be achieved with relative ease due to the encapsulated nature of these lesions.

Diagnosis: Cavernous hemangioma

<p style="text-align: center;">EPIDEMIOLOGY</p> <ul style="list-style-type: none"> • 4.3% of orbital tumors • Female > Male • 3rd to 6th decade (average age = 40) 	<p style="text-align: center;">SIGNS</p> <ul style="list-style-type: none"> • Unilateral proptosis (may accelerate in pregnancy) • Strabismus • Hypermetropic shift • Choroidal folds • Relative afferent pupillary defect • Visual field defect • Lagophthalmos • Elevated intraocular pressure
<p style="text-align: center;">SYMPTOMS</p> <ul style="list-style-type: none"> • Diplopia • Eyelid fullness • Usually painless • Decreased vision 	<p style="text-align: center;">TREATMENT</p> <ul style="list-style-type: none"> • Observation • Surgical excision if threatening vision

Differential Diagnosis:

- Cavernous hemangioma
- Orbital metastasis
- Thyroid orbitopathy
- Orbital lymphoma
- Hemangiopericytoma

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