AMSER Case of the Month June 2024

HPI: 68 year old male with blurry vision

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Patient Presentation

- HPI: 68-year-old male who presents with blurry vision for approximately 1 year. He recently saw his optometrist who had concern for an intraocular lesion.
- Past Medical History: Multiple malignant melanomas of the skin s/p excision, coronary artery disease, hypertension, and multiple gun shot wounds s/p many abdominal surgeries



Objective Findings

- Physical Exam:
 - HENT: Normocephalic and atraumatic. Normal bilateral tympanic membranes, ear canals, and external ears. No congestion or rhinorrhea. No oropharyngeal exudate
 - Eyes: No scleral icterus, no discharge. EOMI bilaterally. Conjunctivae normal. Pupils are equal, round, and reactive to light.
 - Neuro: Alert and orientated to person, place, and time. Baseline mental status. No cranial nerve deficits. Strength and sensation overall intact.
- CBC w diff: Normal
- CMP: Normal



What Imaging Should We Order?



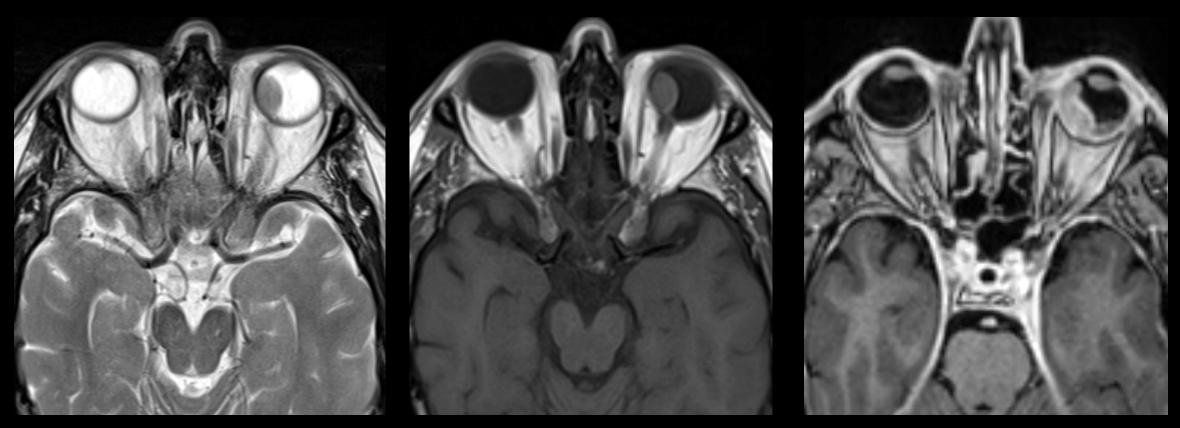
Select the applicable ACR Appropriateness Criteria

Variant 6: Visual loss. Intraocular mass, optic nerve, or pre-chiasm symptoms. Initial imaging.		
Procedure	Appropriateness Category	RRL
MRI orbits without and with IV contrast	Usually Appropriate	0
CT orbits with IV contrast	Usually Appropriate	***
MRI orbits without IV contrast	Usually Appropriate	0
CT orbits without IV contrast	May Be Appropriate	***
MRI head without and with IV contrast	May Be Appropriate	0
CT head with IV contrast	May Be Appropriate	***
MRI head without IV contrast	May Be Appropriate	0
CT head without IV contrast	May Be Appropriate	***
CTA head and neck with IV contrast	May Be Appropriate	***
MRA head and neck without and with IV contrast	May Be Appropriate	0
MRA head and neck without IV contrast	May Be Appropriate	0
Arteriography cervicocerebral	Usually Not Appropriate	***
CT head without and with IV contrast	Usually Not Appropriate	***
CT orbits without and with IV contrast	Usually Not Appropriate	***
X-ray orbit	Usually Not Appropriate	8

This imaging modality was ordered by the physician



Findings (unlabeled)



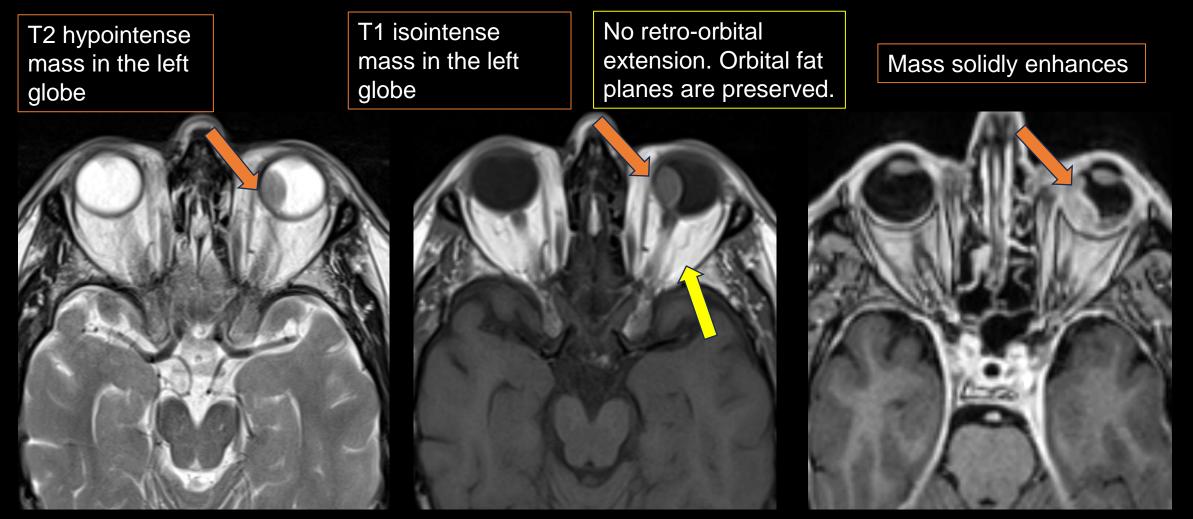
Axial T2

Axial T1 pre-contrast

Axial T1 post-contrast



Findings (labeled)



Axial T1 post-contrast



Axial T2

Axial T1 pre-contrast

Differential Diagnoses

- Metastases, choroidal detachment, uveal neurofibroma, uveal schwannoma, ocular melanoma
- Of Note: Fat, protein, hemorrhage, and **melanin** may be hyperintense on T1 imaging

Final Dx:

Primary Ocular Malignant Melanoma

*thought to be primary because no additional lesions discovered



Case Discussion

Definition

- Ocular melanoma is a rare but deadly malignancy that arises from melanocytes within the uveal tract, conjunctiva, eyelid, and orbit.
- Uveal melanoma is the most common type

Etiology/Epidemiology

- Risk factors include sun exposure, tanning, light skin/eye color, atypical cutaneous nevi, iris nevi, and freckles
- Multiple different genetic mutations have been associated with uveal melanoma although no direct causes
- Most common primary intraocular malignancy
- Primarily seen in the white population
- Incidence increases with age



Case Discussion

Clinical Presentation

- Dependent on location.
 - Choroidal tumors can cause retinal detachment
 - Conjunctival melanomas can present as an elevated brown nodule
 - Ciliary body tumors can cause lens displacement and disturbances in accommodation
 - Iris melanomas can distort the pupil and cause cataracts and glaucoma
- Commonly asymptomatic at diagnosis
- Common symptoms include blurry vision, visual field defect, flashing lights, redness, irritations, pain, and a pressure-like sensation
- Monitoring for metastasis should be performed regularly regardless of stage and treatment



Case Discussion

• Pathology

- Primarily believed to be due to oxidative damage
- Histologically, mixed melanoma containing both spindle and epithelioid cells is the most common subtype.
 - This case was confirmed by pathology to be uveal melanoma, epithelioid type with evidence of extrascleral spread

Treatment and Prognosis

- Treatment options based on tumor size include laser photocoagulation, transpupillary thermotherapy, plaque radiation therapy, particle beam radiotherapy, local surgical resection, and enucleation
- Poor prognosis factors: age > 60 years, larger tumors, anterior location within the globe, epithelioid cells, and extraocular extension
- High risk of distant metastasis: most commonly liver, lung, and bone
- 50% mortality rate despite treatment advances



References:

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- Jha P, Hacking C, Yap J, et al. Primary uveal malignant melanoma. Reference article, Radiopaedia.org. Accessed on 21 Mar 2024. <u>https://doi.org/10.53347/rID-8552</u>.
- Papayiannis V, Tsaousis KT, Kouskouras CA, Haritanti A, Diakonis VF, Tsinopoulos IT. Investigation into the quantitative and qualitative characteristics of choroidal melanoma through magnetic resonance imaging and B-scan ultrasound. Clinical Ophthalmology. 2017; Volume 11. 1557-1564. doi: 10.2147/OPTH.S130009.
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