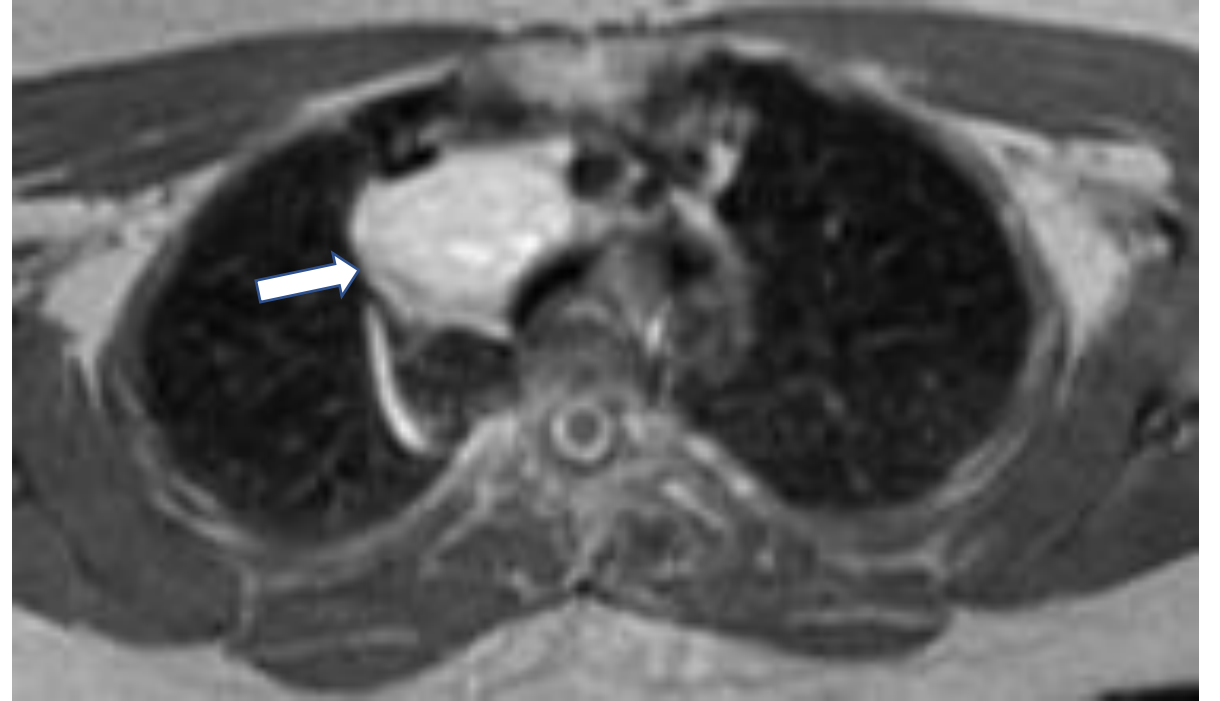
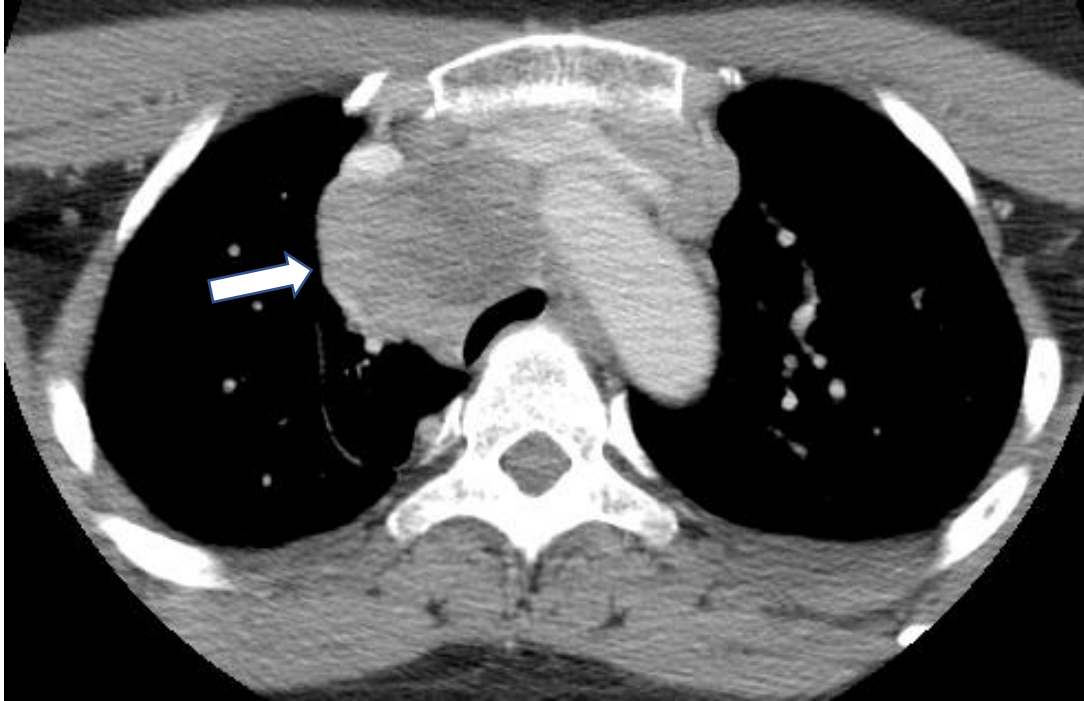


# Choroidal neuroendocrine metastases

Nir Stanietzky, MD; Inessa Goldman, MD

# History

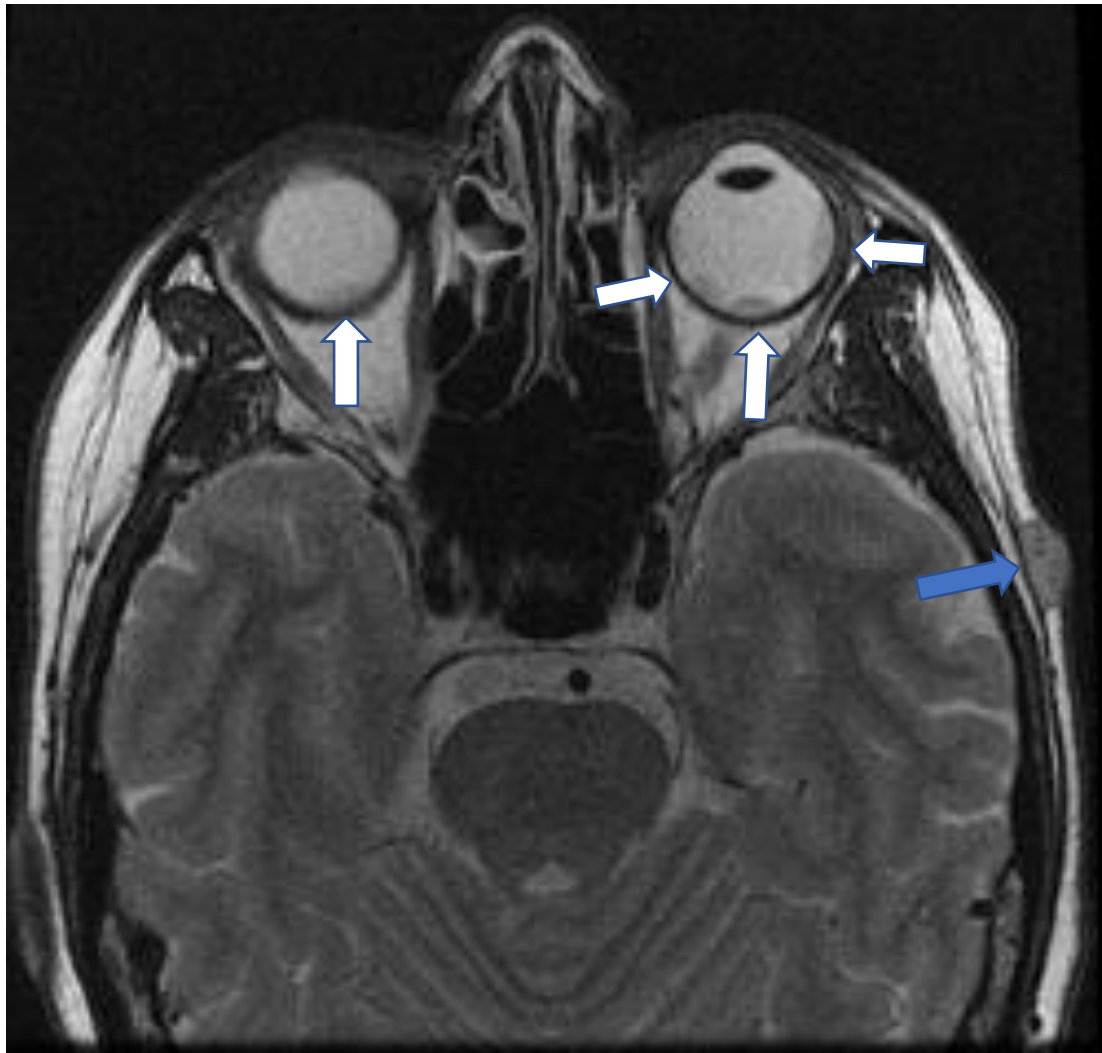
- A 49 year old female presented with a firm, rapidly growing subcutaneous nodule on the underside of her forearm.
- An excisional biopsy was performed approximately one month later.
- The mass was found to be neoplastic and a full diagnostic workup was performed including a CT scan and MRI of the chest, abdomen and pelvis.



A contrast enhanced axial CT scan of the chest demonstrates a large heterogeneous enhancing multicompartmental mediastinal mass (white arrows). This is also seen on a representative axial T2 weighted SSFSE MRI image.

# Clinical Course

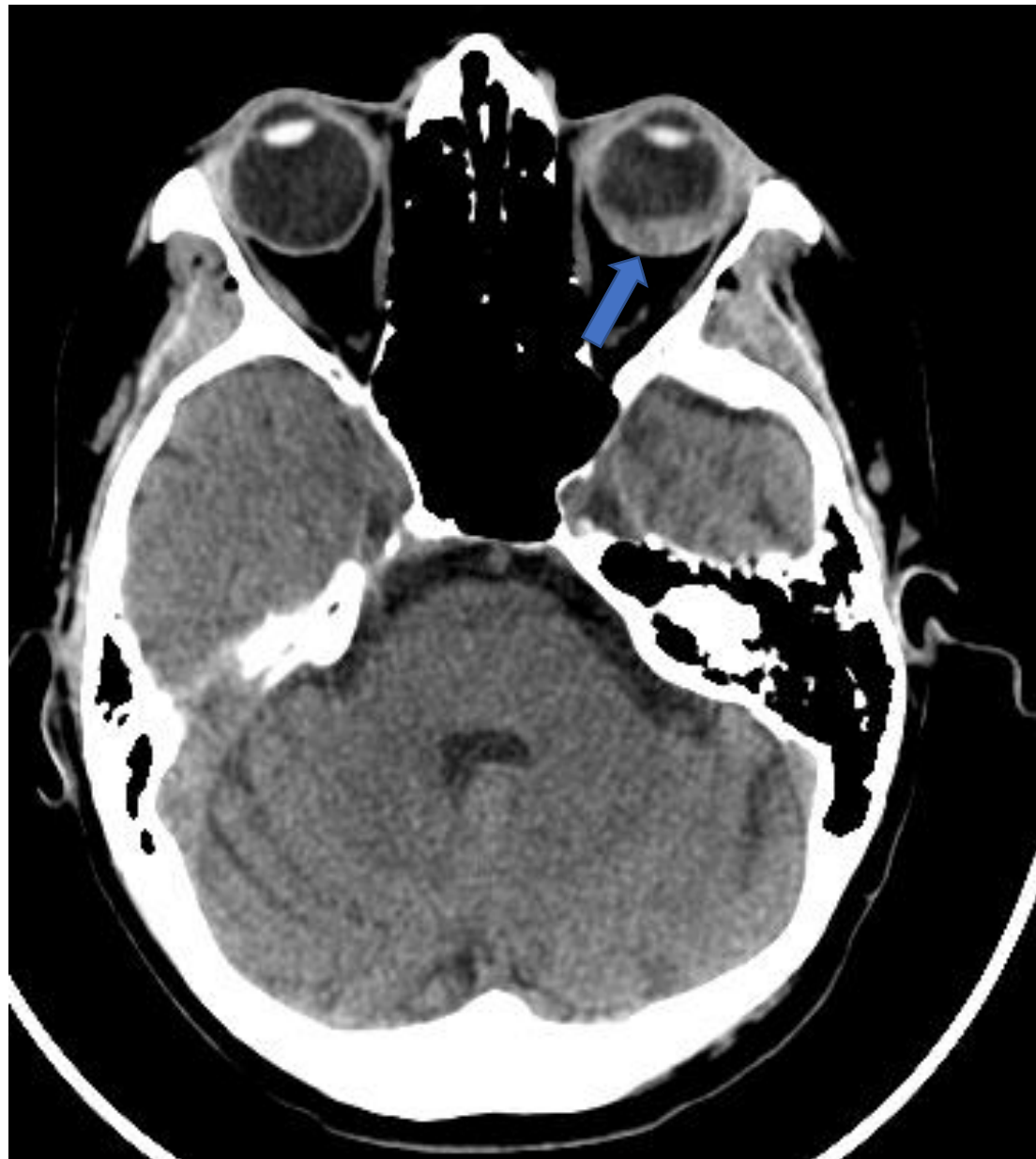
- The primary site was felt to be the large mediastinal mass.
- A biopsy of a suspicious left supraclavicular lymph node demonstrated a high-grade neuroendocrine neoplasm metastasis.
- The patient was treated with Avastin, Sandostatin, Temodar.
- She progressed and her treatment was modified to Xeloda and Temodar
- Approximately two years later, the patient developed decreased vision in her left eye and presented for ophthalmologic assessment.
- A mass was seen on fundoscopy, and an MRI was ordered.



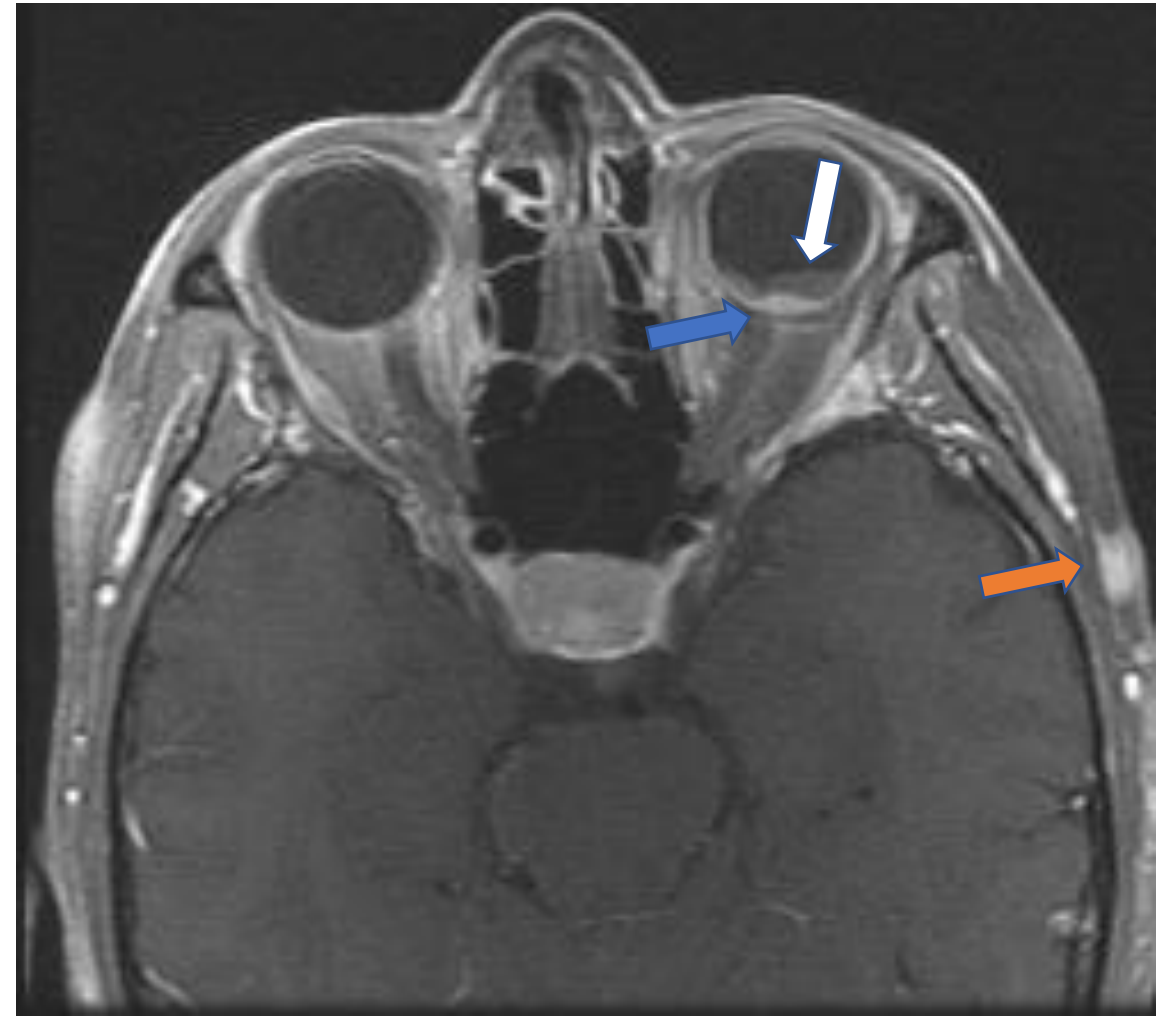
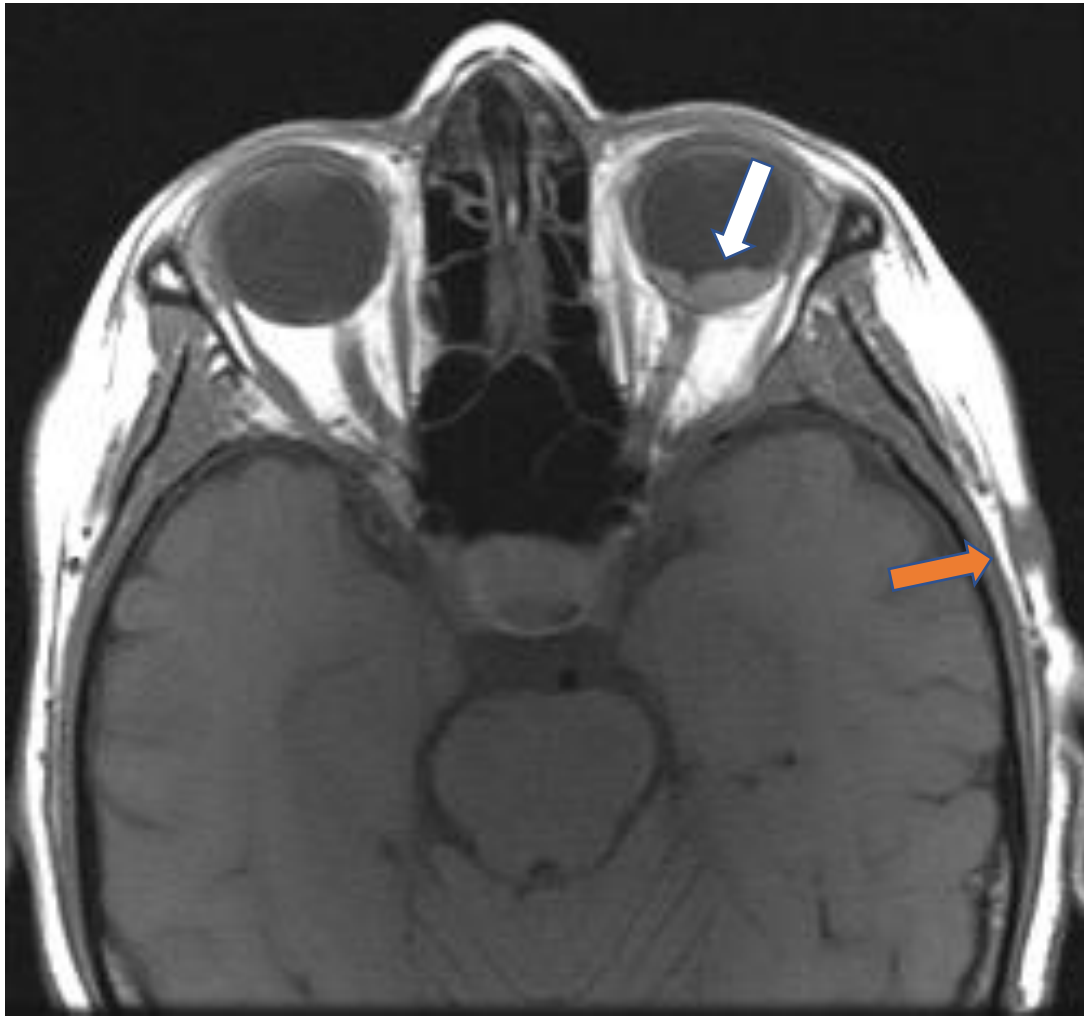
Axial T2 and post contrast sagittal T1 weighted images demonstrate bilateral enhancing choroidal lenticular masses (white arrows) presumed to be metastatic in nature. Also seen is a left temporal skin metastasis (blue arrow).

# Clinical Course

- She was treated with external beam radiation therapy.
- One month later, the patient developed an acute severe headache.
- At the same time, she noticed that her left visual field seemed as if “a curtain was coming down, associated with flashing lights.”
- She presented to the emergency room. An emergent non-contrast CT scan and subsequently a contrast enhanced MRI were performed.



Non contrast axial CT scan of the head demonstrates a new **hyperdense crescentic** left retinal hemorrhage overlying the area of the previously documented choroidal metastases (blue arrow).

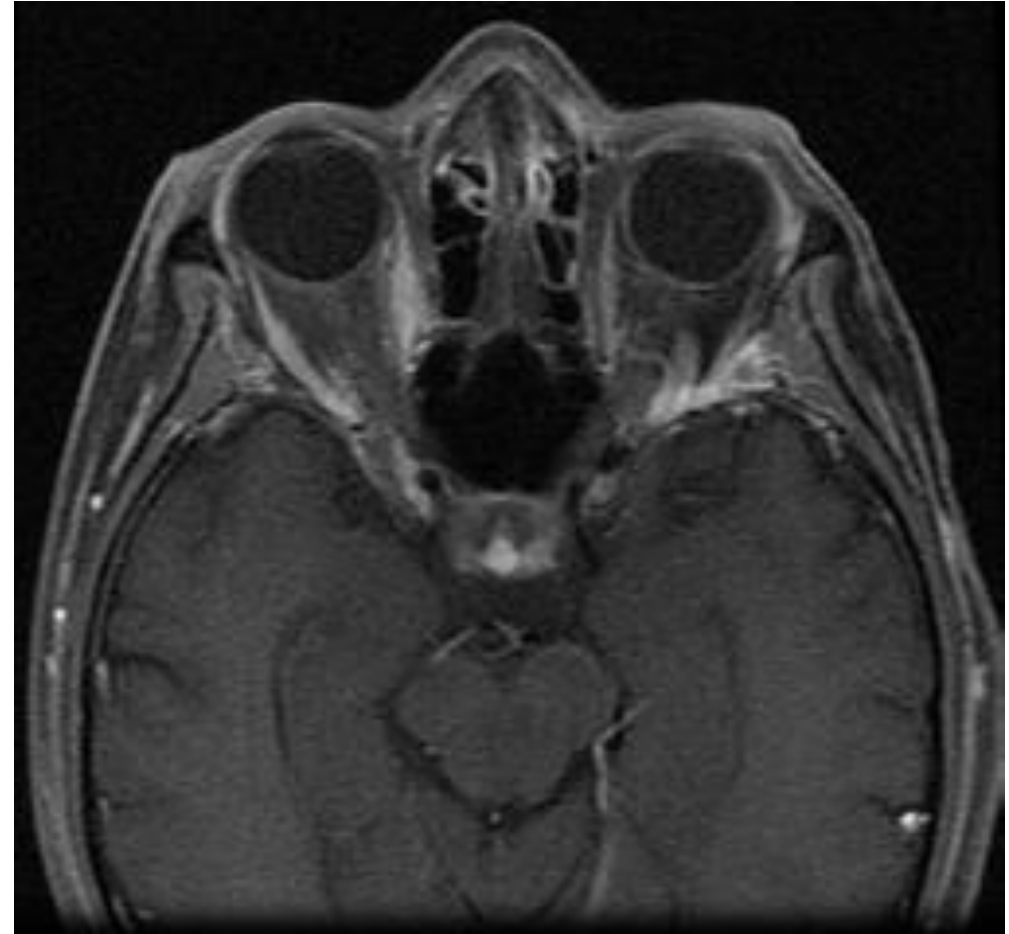
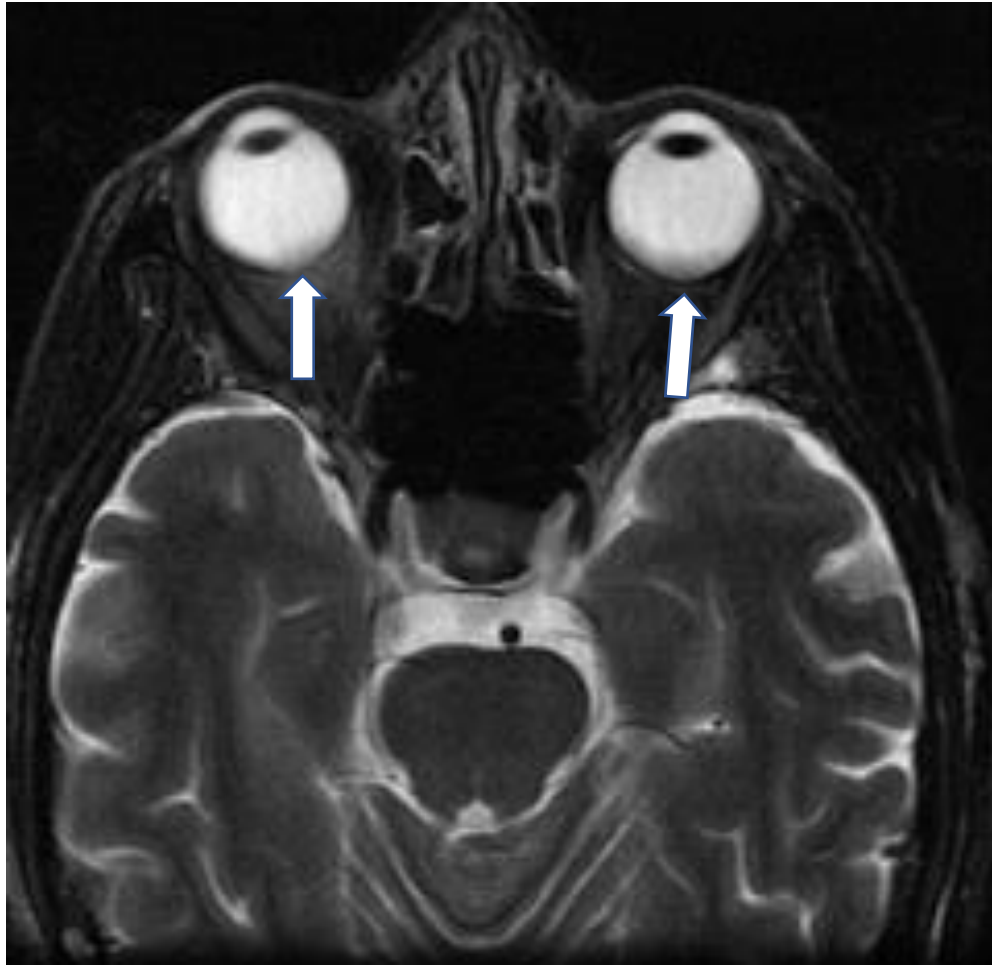


Pre- and post-gadolinium T1 weighted axial MRI of the orbits demonstrate an enhancing left choroidal metastasis (blue arrow) with surrounding hemorrhage and retinal detachment (white arrows). A left temporal enhancing subcutaneous metastasis is also seen (orange arrow).



# Clinical Course

- She underwent Indocyanine Green-Enhanced Transpupillary Thermotherapy to the left eye and continued external beam radiotherapy.
- Her symptoms significantly improved, and a repeat MRI was performed three months later.
- Eventually, the patient developed progressive multifocal metastatic disease to the skin, liver, bones and peritoneum and expired approximately one year later.



Three months later, T2 and post contrast T1 weighted MRI images shows residual bilateral choroidal metastases (white arrows), but resolution of the left sided retinal hemorrhage.

# Epidemiology

- The vast majority of uveal tumors are choroidal melanomas, however rarely a choroidal mass may represent metastases from a neuroendocrine tumor.
- Neuroendocrine tumor ranks sixth in frequency (2%) for choroidal metastases. Breast cancer and lung cancer are two most common primary sites (~44% and 25%, respectively).
- The 5-year survival rate for all metastases is poor at 23%, but lung neuroendocrine metastasis demonstrate the most favorable 5-year survival rate at 92%.
- Choroidal metastases are generally highly vascular tumors, like choroidal hemangioma.

# Clinical Features

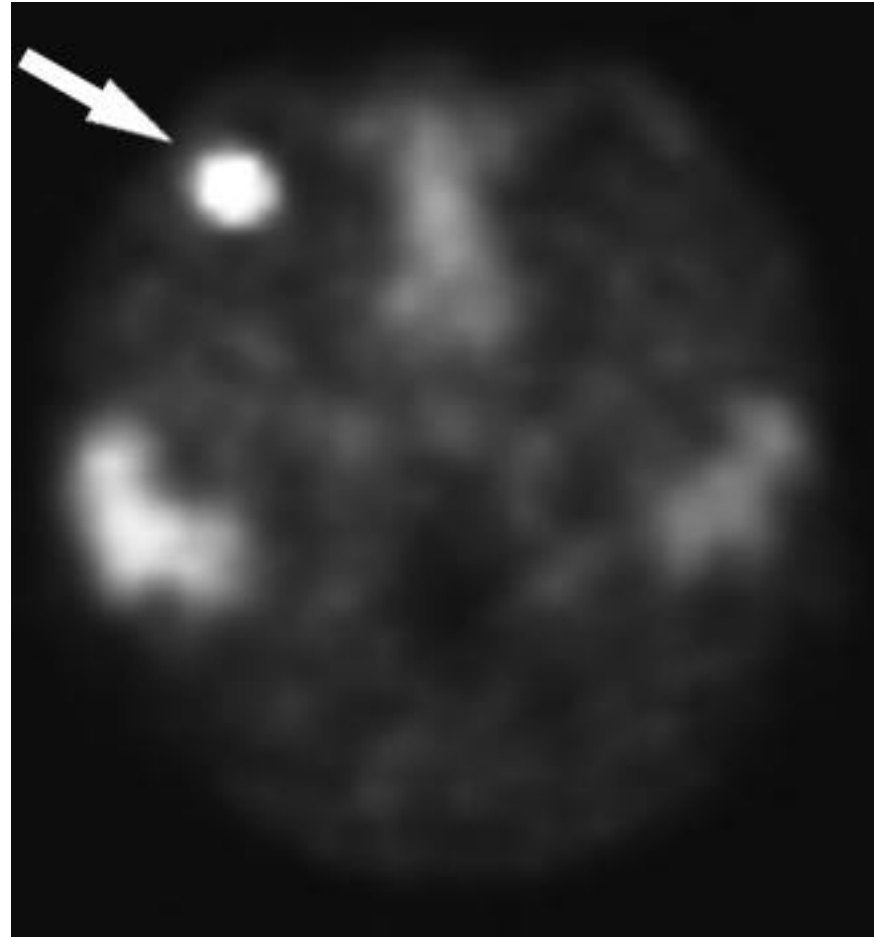
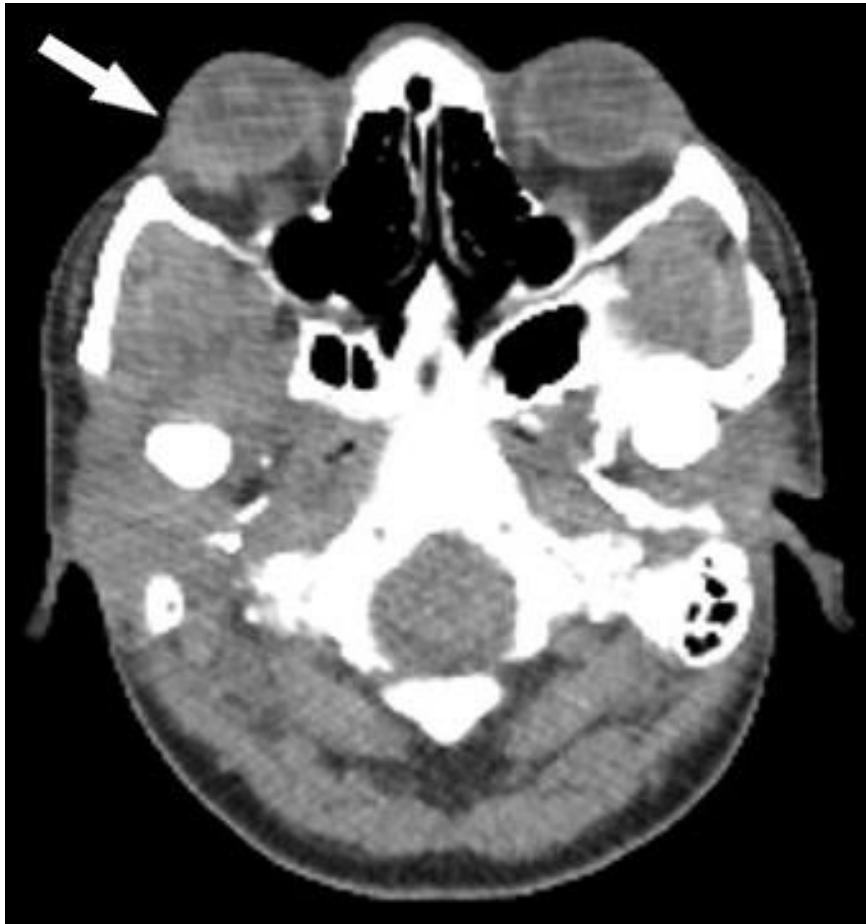
- Choroidal neuroendocrine metastases usually present in patients with widespread metastatic disease.
- They are most often non-melanotic, presenting as orange or orange-yellow in color.
- It is exceedingly rare to develop metastases from pigmented tumors of nonmelanocytic derivation, but this can happen with neuroendocrine tumors.
- On fundoscopic exam, the pigmented tumors are often misdiagnosed as a choroidal melanoma
- If the diagnosis is not suspected, pigmented tumors present a significant diagnostic challenge to ocular cytopathologists due to presence of melanin.

# Clinical Features

- Immunohistochemical analysis needs to be performed when atypical features are seen in a pigmented choroidal tumor thought to be a melanoma. Immunoreactivity for chromogranin, synaptophysin, cytokeratin, and calcitonin and detection of dense-core neurosecretory vesicles on electron microscopy helps to establish neuroendocrine origin of these choroid metastases.
- Melanomas classically present as a mushroom-shaped choroidal mass due to the ability of these tumors to erupt through Bruch's membrane.
- Choroidal metastases, in contrast, rarely adopt this growth pattern, but rather a collar-button configuration on close inspection.

# Imaging

- On ocular ultrasonography, neuroendocrine metastasis is usually echogenic, while melanoma is typically echolucent.
- They are seen as enhancing choroidal masses on CT scan and MRI.
- Due to the hypervascularity of these lesions, they can present with choroidal hemorrhage and detachment.
- These are now seen much more commonly with somatostatin receptor analog imaging with one study demonstrating metastases to the eye and orbit in up to 15% of patients with late-stage disease.



Images from a *Ga68 DOTATATE PET/CT* from a different patient demonstrate presence of choroidal somatostatin-receptor bearing neuroendocrine metastases (white arrow).

# Diagnosis/Treatment

- Fine needle aspiration can be performed but carries the risk of hemorrhage and possible blindness due the hypervascularity of the tumor.
- They can be treated with external beam radiotherapy, plaque radiotherapy, laser photocoagulation, anti-vascular endothelial growth factor therapy, and photodynamic therapy.
- In patients with widespread disease, labeled radiopharmaceutical analogs, most commonly with Yttrium-90 and Lutetium-177, can be used.



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