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Case of Optic Nerve Sheath Meningioma

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Abstract: ***A 32 year old female, known hypothyroid on treatment and no other comorbidities had come with the complaints of reduced vision in the left eye. Opthalmoscopic examination showed optic disc swelling. There was mild proptosis. On computed tomography of brain, there was thickening around the left optic nerve with calcifications. Brain parenchyma was normal. On MRI plain and contrast, there was thickening around the left optic nerve which showed homogenous enhancement post gadolinium contrast administration. This thickening of the optic nerve sheath caused compression and atrophy of the left optic nerve, resulting in the patient's complaints of reduced vision on the left side. Brain parenchyma was normal. There was no intra-parenchymal extension of the lesion. Orbital muscles were normal.***

Keywords: Optic nerve sheath meningioma - benign optic nerve glioma (mostly in children) and orbital pseudotumour (inflammatory) are the two closest differentials

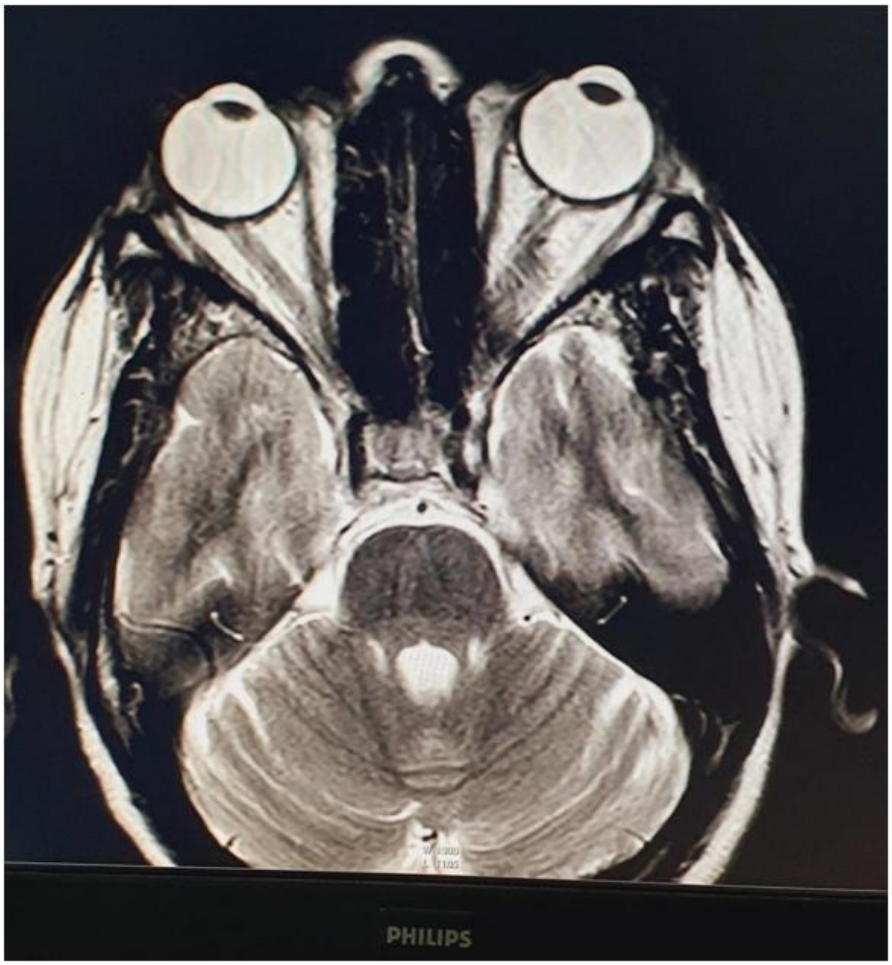
1. History

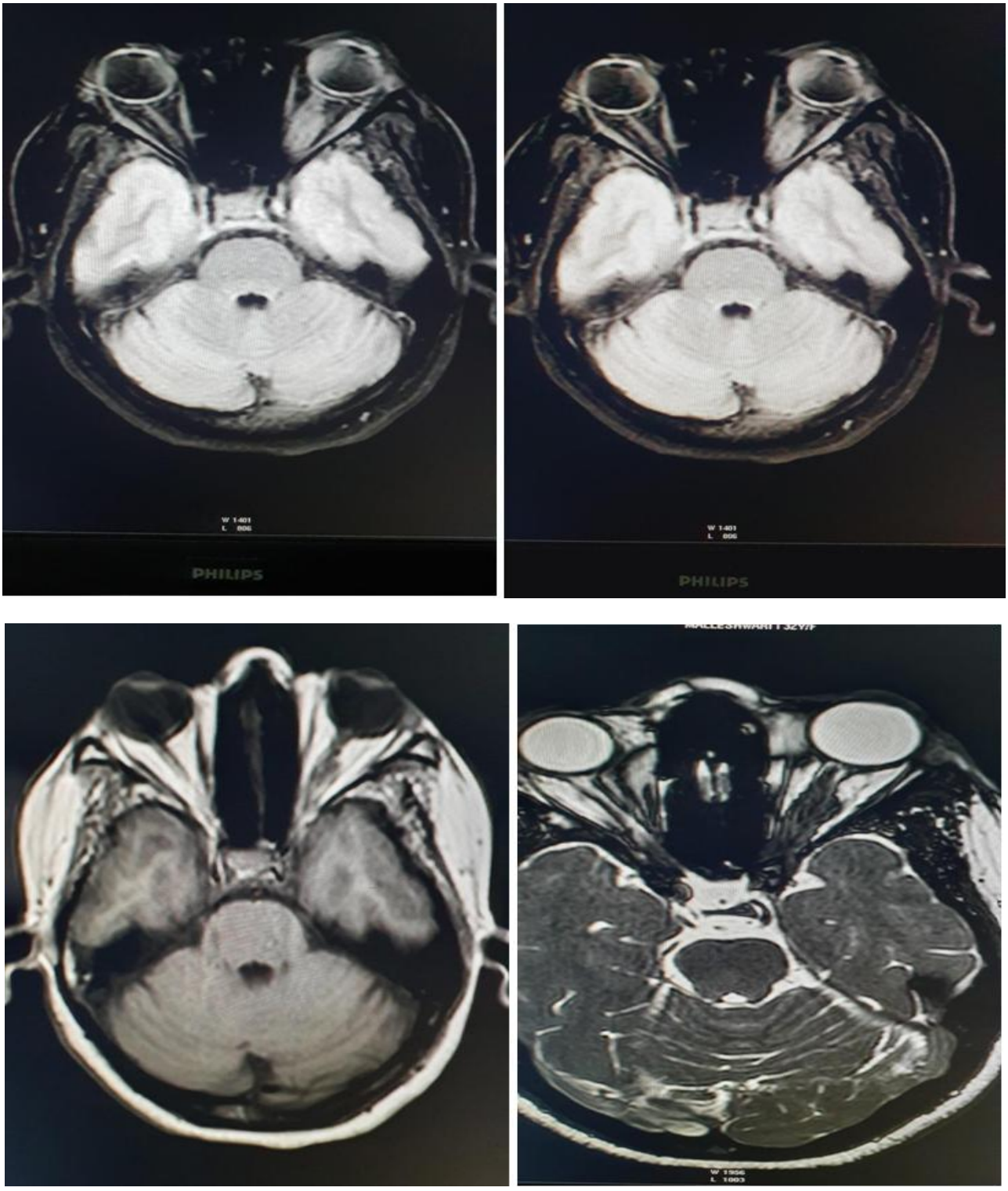
- A 32 year old female, known hypothyroid on regular medication was referred to our department for MRI of orbits in view of reduced vision in left eye.
- The patient was previously healthy with no other complaints, she had mild hypothyroidism for which she was under regular treatment and her T3, T4 and TSH levels were within normal limits.

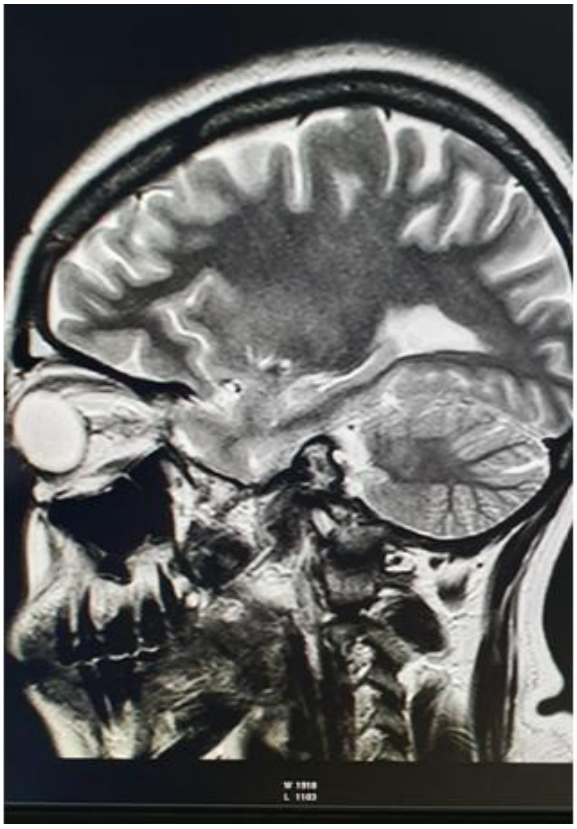
2. Clinical Examination

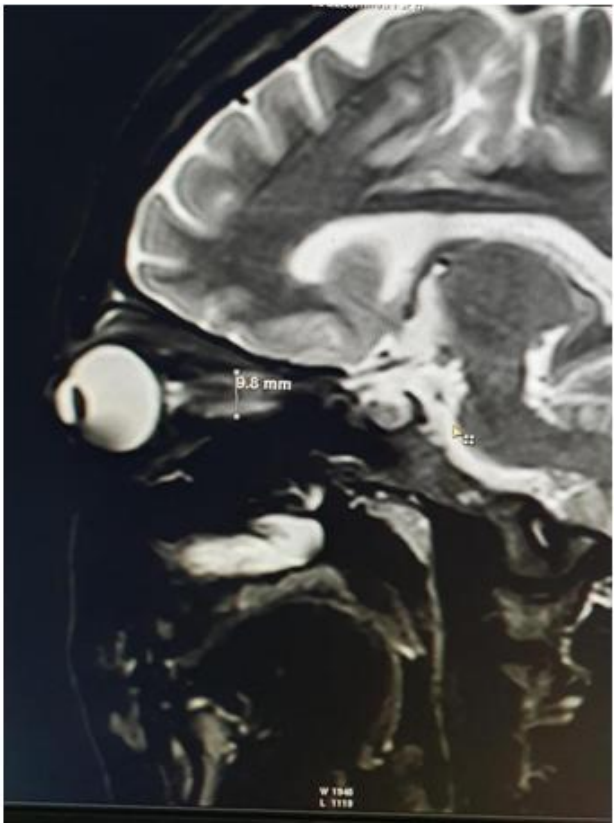
- Opthalmoscopic examination conducted at the Department of ophthalmology, at our institution revealed optic disc swelling.
- There was mild proptosis and lower lidedema.

MRI



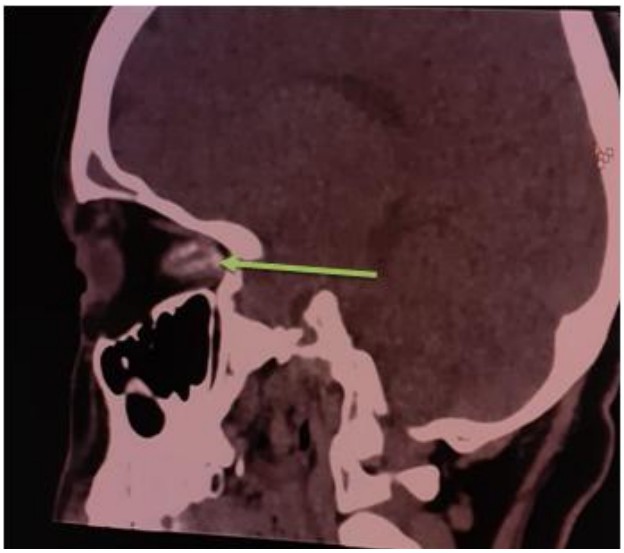


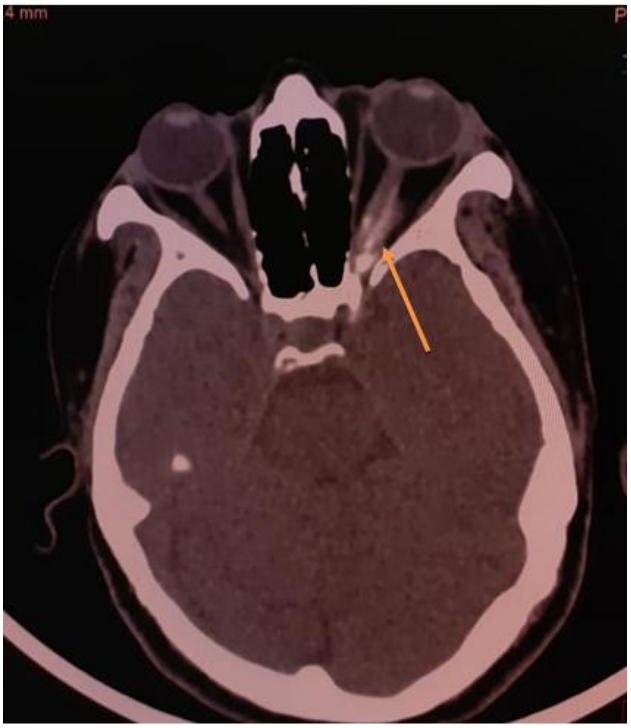




Plain MRI on T1 and T2 (in coronal, axial and sagittal views) sequences revealed thickening around the left optic nerve suggestive of nerve sheath thickening which caused compression effect on the optic nerve, ultimately causing its atrophy

A non – contrast computed tomography was done which showed calcification around the nerve, which is again in favor of optic nerve sheath meningioma

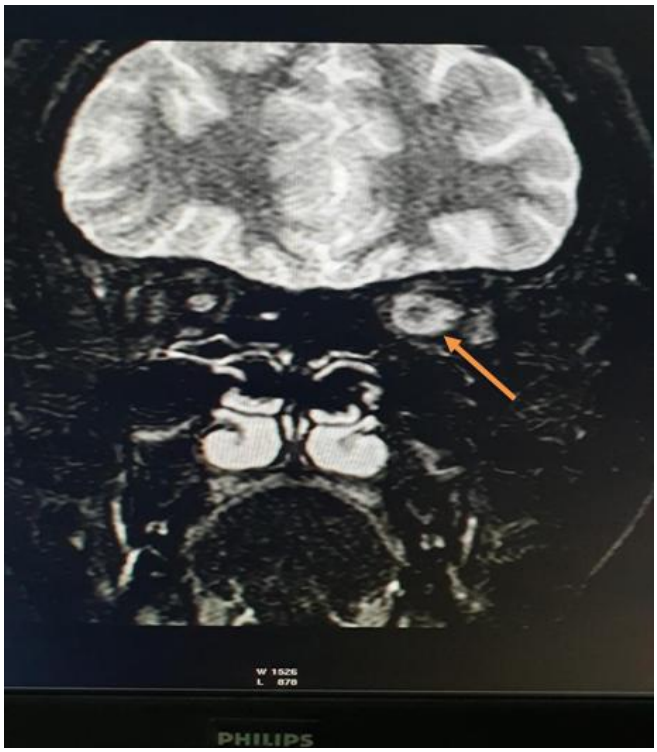


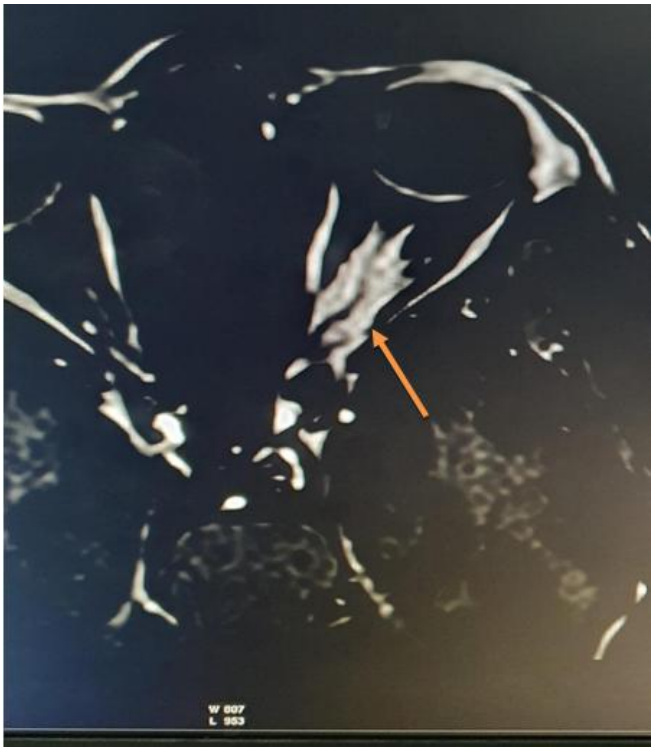


Following MRI an academic NCCT of the orbits was performed which showed calcifications along the sheath.

The left optic nerve was a trophic. Mild proptosis of the left eye.

Brain parenchyma was normal.





Post Contrast

3. MRI with Contrast Findings

- TECHNIQUE: T1 fats at/ T2 fats at axials
  STIR oblique sagittals
  T2fatsat/T1fats at coronals
- E/OT1, T2 iso intense to grey mater lesion noted around the left optic nerve, causing compression of the nerve.
- Post contrast the lesion is showing vivid enhancement with sparing of the nerve resulting in doughnut sign on coronals and tram track signonaxials–S/O optic nerve sheath meningioma

4. Discussion

- Optic nerve sheath meningiomas are benign tumors arising from the arachnoid cap cells of the optic nerve sheath and represent~ 20% of all orbital meningiomas, the majority of which are direct extensions from intracranial meningiomas.
- These tumors typically appear as masses within the optic nerve, iso intense to grey matter on both T1 and T2 weighted imaging, demonstrating vivid enhancement which clashes with the non-enhancing optic nerve
- ("tram-track sign"on axials or
- "non-enhancing dot sign “or doughnut sign on coronals).

Epidemiology

- Account for approximately a third of all optic nerve neoplasms; optic nerve gliomas are the most common entity.
- Unlike optic nerve gliomas which occur primarily in children, optic nerve meningiomas are usually seen in adults (meanage at presentation 40 years)
- Similar to meningiomas else where there is a female predilection.

- The vast majority of cases are sporadic, although patients with neurofibromatosis type II (NF2) are at increased risk

Clinical Presentation

Visualloss	Proptosis
Present to a greater or lesser degree in 95% of cases	Present to a greater or lesser degree in 60-90% of cases
Painless and progressive	More pronounced with anterior lesions near the globe
More pronounced with lesions that affect the orbital apex	Occurs later, as a tumor enlarges
Exacerbated during pregnancy	

- May be exacerbated by associated hyperostosis
- These lesions are almost always unilateral, with the exception of NF2 patients who may develop bilateral tumors.
- Occasionally a unilateral tumor will grow posteriorly, across the chiasm and along the contra lateral nerve.
- A particular clinical presentation is worth mentioning due to the potential pitfall
- An intra canalicular lesion may cause marked visual loss with a very small tumor and clinical presentation may be similar to optic neuritis. As these patients are often middle-aged women, the diagnosis of multiple sclerosis may be suspected. Thus in patients being imaged for 'optic neuritis' careful inspection of the optic canals is important

Pathology

- Optic nerve meningiomas arise from the arachnoid cap cells of the optic nerve sheath, and as such are on the inside of the dura
- The tumor extends through the over lying dura and typically has a smooth or somewhat lobulated contour.
- The optic nerve, which is usually circumferentially encased, gradually atrophies due to compression.
- The most common type of meningioma histologically is the meningothelial variety

Radiographic Features

- Meningiomas of the optic nerve sheath have the same imaging characteristics as meningiomas elsewhere. The morphology of the tumor is variable:
- Tubular:65%
- Exophytic:25%
- Fusiform:10%

CT

- The tumor is usually is attenuating to the optic nerve on non-contrast studies, although calcification is sometimes seen.
- On axial or oblique sagittal imaging the enhancing tumor surrounding the non-enhancing optic nerve results in the so-called tram-track sign. This is most evident in tumors with tubular growth pattern.
- On coronal imaging, the tumor appears as a cuff of enhancing tumor around a central non-enhancing dot (optic nerve) giving a doughnut appearance (doughnut sign).
- Tumor extending in to the optic canal may lead to canal widening, or alternatively hyperostosis

MRI

- Appearances on MRI are similar to those on CT with a greater ability to delineate posterior extension.
- Imaging should include thin axial and coronal (+/- sagittal) T1, fat-suppressed T2 and fat- suppressed post contrast T1 sequences.
- T1: isointense to somewhat hypo intense compared to the optic nerve
- T1C+(Gd): homogeneous enhancement
- T2: isointense to some what hyperintense compared to the optic nerve
- Careful examination of the orbital apex and optic nerve canals is essential if small intracanalicular tumors are to be identified

Differential Diagnosis

- Optic nerve glioma
- Orbital pseudo-tumor
- Orbital lymphoma/ leukemia
- Orbital Metastases
- Sarcoidosis

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