

## Case Report

### CONTRALATERAL OPTIC NEUROPATHY IN SPHENOID WING MENINGIOMA WITH MIDBRAIN COMPRESSION

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#### ABSTRACT

A 38-year-old lady presented with progressively worsened binocular diplopia for 2 months associated with periorbital pain, headache, nausea and right sided tinnitus. Ophthalmic examination revealed Left Relative Afferent Pupillary Defect (RAPD) with impaired optic nerve function and right 6<sup>th</sup> cranial nerve palsy. Fundus examination showed bilateral optic disc swelling, more in the left eye. Humphrey Visual field showed left superior quadrant field defect. MRI Scan revealed right sphenoid meningioma with midline shift causing compression of the adjacent ventricle. She underwent preoperative angioembolization of the tumour followed by right craniotomy and tumour excision. Post-operatively, the diplopia, RAPD, optic disc swelling and visual field defect resolved. Optic neuropathy may be worst on the contralateral side of a space-occupying lesion despite bilateral optic disc swelling.

#### INTRODUCTION

Papilloedema is a medical emergency as it indicates raised intracranial pressure (ICP). A raised ICP may result in compressive optic neuropathy, particularly causing Relative Afferent Pupillary Defect (RAPD) on the ipsilateral side of the more severe optic disc swelling, alongside other compressive features affecting cranial nerve involvement, commonly the 6<sup>th</sup> cranial nerve. Contralateral RAPD may however be seen in optic tract lesions due to decussation of the optic nerve fibres at the optic chiasm. We report a case of a patient with a right sphenoid wing meningioma with increased ICP and an RAPD on the contralateral side of the tumour.

#### CASE REPORT

A 38-year-old lady with no known medical illness presented with binocular horizontal diplopia for 2 months. It was initially intermittent but worsened to persist throughout the day. The symptom was relieved when she turned her head towards the left side. There was no blurring of vision, central scotoma or metamorphopsia. The diplopia was associated with a right-sided tinnitus and headache which worsened upon waking up from sleep or when bending forward. She also had periorbital pain which was aggravated on eye movement. She had nausea but no vomiting for 1 month. There was no other significant neurological complaint.

Ophthalmic examination revealed best corrected visual acuity of 20/60 in both eyes. There was presence of Relative Afferent Pupillary Defect (RAPD) and reduced optic nerve function on the left eye (LE). Further examination showed limitation in the right lateral gaze consistent with right 6<sup>th</sup> cranial nerve palsy evidenced on Hess chart. Fundus examination showed bilateral optic disc swelling, more profound in the LE. Humphrey Visual field (HVF) showed incongruous left homonymous hemianopia with an enlarged blind spot in both eyes. Other neurological examination was unremarkable.

Magnetic resonance imaging of the brain revealed a well-circumscribed extra-axial mass at the right middle cranial fossa established by the presence of a CSF cleft and cortical buckling (Figure 1), suggesting a sphenoid wing meningioma. The lesion exerted a mass effect to its surrounding structures such as right temporal and frontal lobes and displacing the right middle cerebral artery. There was also effacement of the right lateral ventricle and significant midline shift with uncal herniation which explained the symptoms of elevated intracranial pressure.

The patient underwent preoperative angioembolization of the tumour followed by right craniotomy and tumour excision. Histopathology examination showed neoplastic meningothelial cells arranged in whorled storiform patterns and syncytial-like lobules set in collagen rich matrix (Figure 2a). The tumour cells exhibit mild nuclear

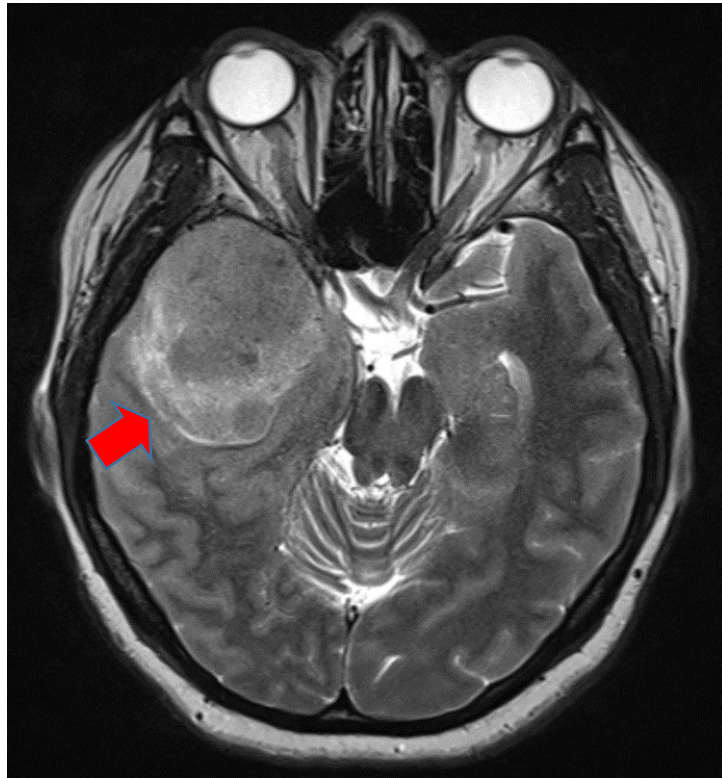


Figure 1: Axial view of T1-weighted MRI imaging showing well circumscribed extra-axial mass at the right middle cranial fossa measuring 5.4cm x 5.6cm x 6.3cm (AP x W x CC). There is midline shift to the left with uncus herniation and displacement of the right middle cerebral artery and midbrain. Bilateral globes are normal with no optic nerve compression.

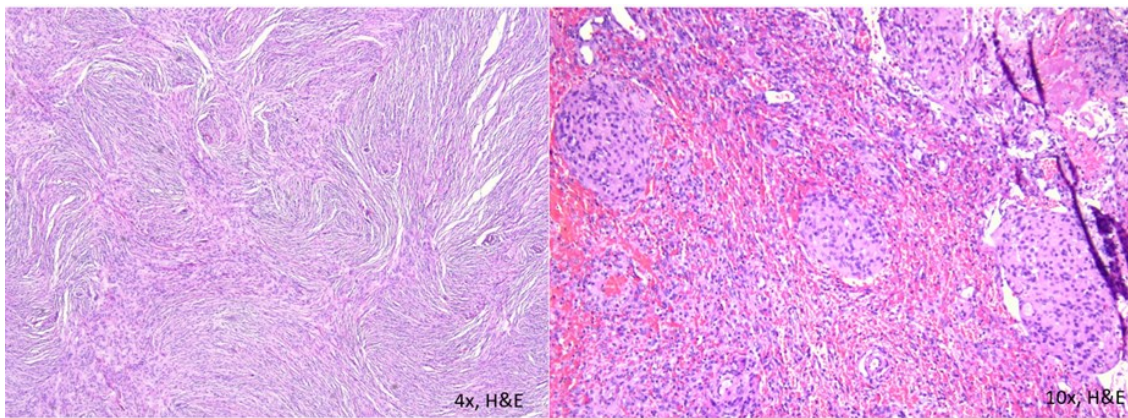


Figure 2a: H&E stain of the tumor tissue showing the tumour composed of neoplastic meningotheelial cells arranged in whorled storiform patterns and syncytial-like lobules set in collagen-rich matrix depicting meningioma

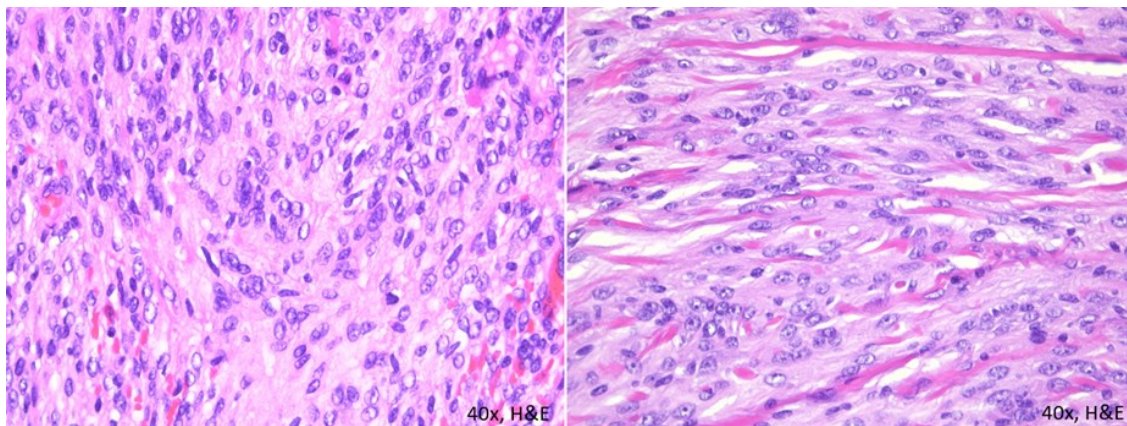


Figure 2b: H&E stain showing the tumour cells exhibiting mild nuclear atypia, round to oval, vesicular nuclei, nuclear haloes, pseudoinclusions, inconspicuous nucleoli and indistinct cytoplasmic borders. No anaplastic cells were observed.

atypia, round to oval vesicular nuclei, nuclear haloes, pseudoinclusions, inconspicuous nucleoli and indistinct cytoplasmic borders (Figure 2b). A diagnosis of sphenoid meningioma was confirmed.

Six weeks post-operatively, the patient recovered completely with no ocular complaints. Vision improved to 20/20 with no diplopia and complete eye movements in all gaze. There was also complete resolution of the left RAPD. Bilateral fundus examination showed resolving bilateral optic disc swelling (Figure 3). HVF showed normal VF with no defects (Figure 4).

## DISCUSSION

Ophthalmic manifestations of space occupying lesions are indeed a useful aid in localisation of the lesion. While neurological symptoms may often be non-specific, ocular findings including mapping of visual fields can assist in correctly determining the position of the tumour in the brain.

RAPD is usually present when there is optic nerve dysfunction causing abnormal light transmission in the afferent pupillary pathway. It is however most common due to a pre-chiasmal pathology causing the defect to be detected on the same side of the insult. A contralateral RAPD can also happen, albeit less frequently. This occurs in optic tract lesions due to decussation of the optic nerve fibres at the optic chiasm whereby 53% of fibres receives input from the contralateral nasal retina. This is attributed by the presence of higher photoreceptor density in the nasal than the temporal retina. Because of this asymmetric distribution of photoreceptors and the ratio of crossed to uncrossed fibers in the chiasm, an RAPD can also result from contralateral optic tract and midbrain lesions [1]. The lesion is more suggestive to be at the optic tract when it is accompanied with complete homonymous hemianopia. The pupillary defect will be present on the side opposite to the lesion, resulting from damage to afferent nasal fibers that cross at the optic chiasm which will then continue on the optic tract and subsequently synapse at the pretectal

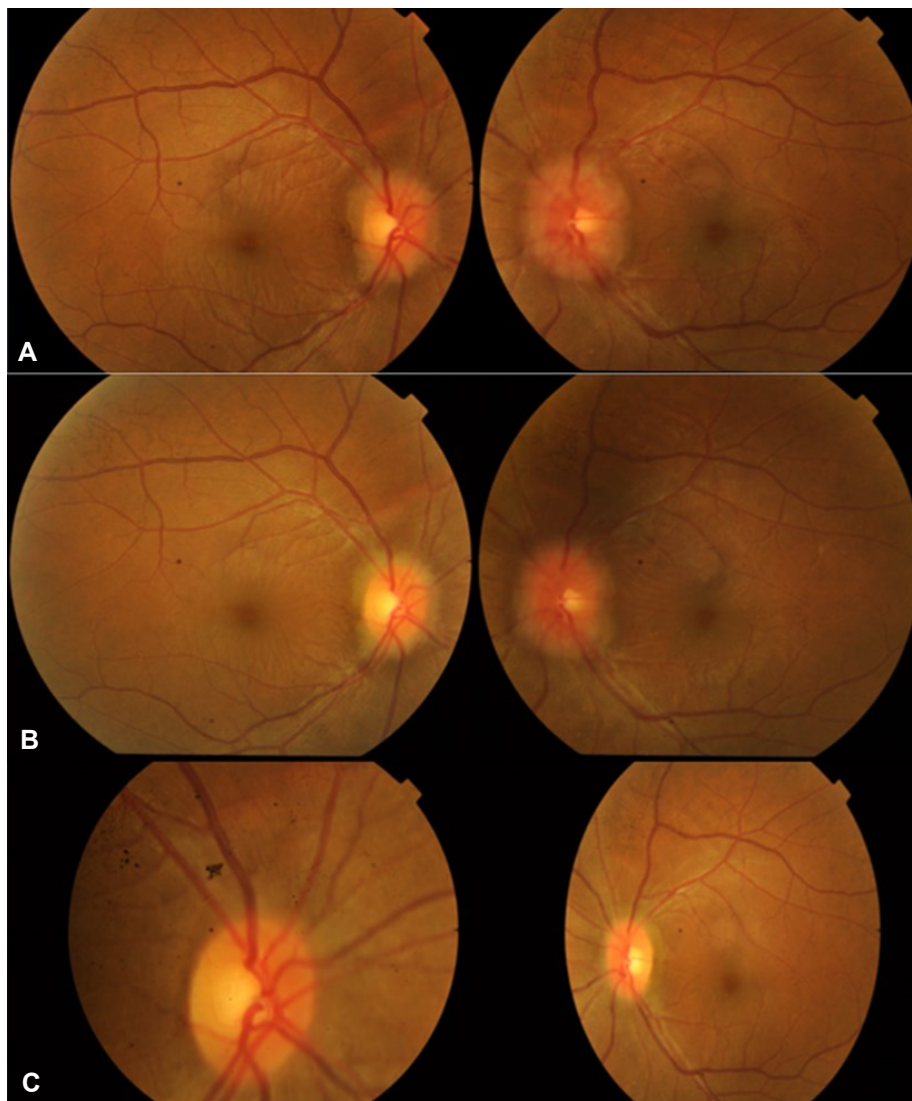


Figure 3: (A) Coloured fundus photograph showing bilateral optic disc swelling at presentation. (B) One week post tumour excision showed reduction in optic disc swelling with clearer disc margins. (C) Six weeks post tumour excision showing complete resolution of the optic disc swelling.

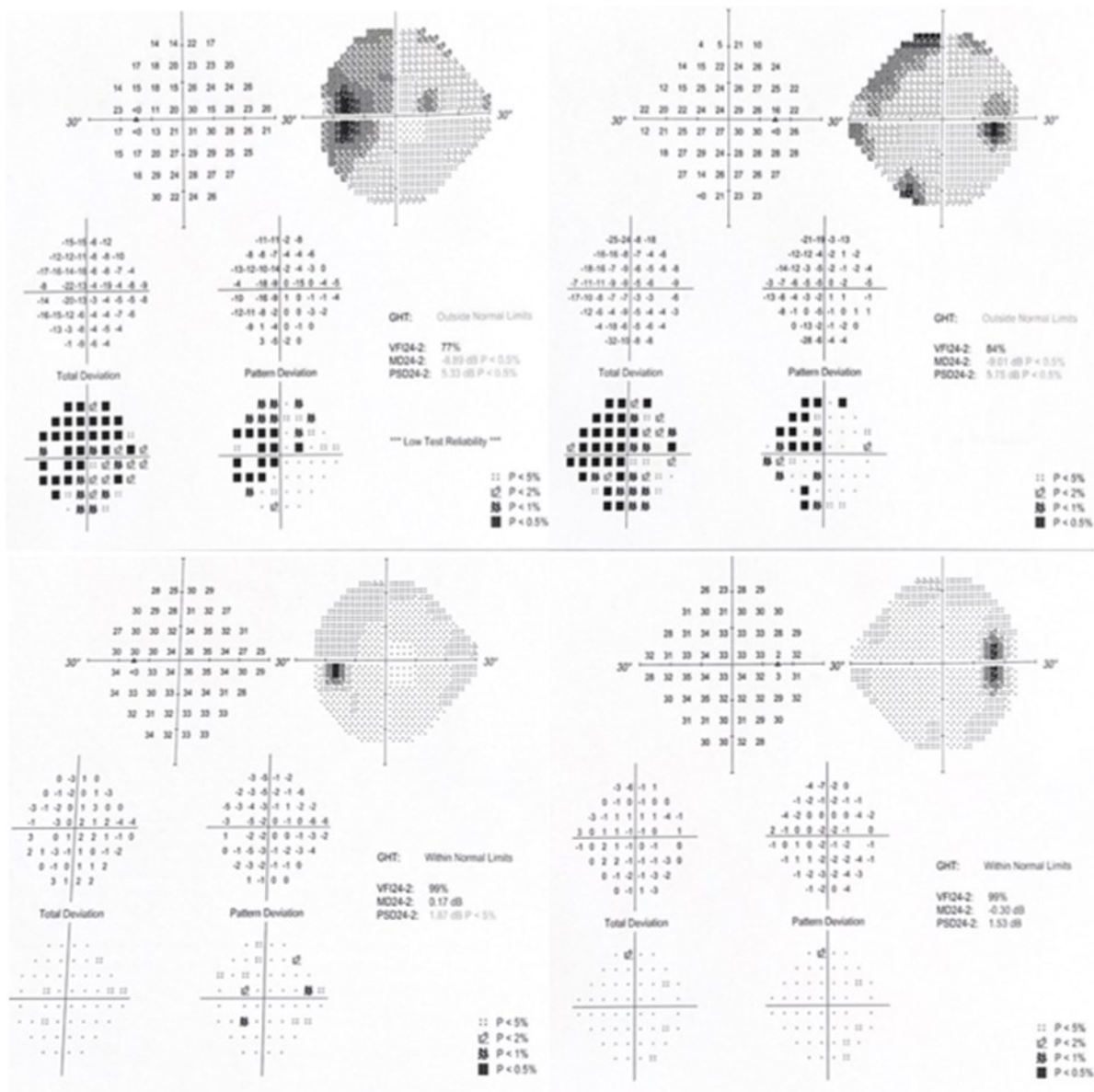


Figure 4: (Top) 24-2 Humphrey visual field on presentation showing incomplete left homonymous hemianopia with (Bottom) resolution of visual field defect 6 weeks post tumour excision.

nuclei, before partly decussating to synapse at the Edinger-Westphal nuclei as part of the afferent pupillary light reflex pathway [2].

The presence of RAPD can also distinguish between an optic tract lesion or damage posterior to the lateral geniculate nucleus as there will be a band-shaped atrophy of the contralateral optic disc in optic tract lesion [3]. In certain cases, RAPD may be seen despite the absence of any afferent visual pathway lesion [4]. This can be further explained as the midbrain contains retinotectal tract that is made up of a small subset of ganglion cells where 10% retinal afferent fibers bypass the lateral geniculate nucleus and is relayed here [1]. This tract will then transverse the superior colliculus and subsequently synapse at the pretectal nucleus near the junction of midbrain and forebrain. From here it projects to the Edinger-Westphal nucleus, the postganglionic ciliary ganglion where impulse is sent to the pupillary constrictors as

well as ciliary muscles. Lesions affecting these fibers that have branched off from the optic tract during their trajectory to the dorsal midbrain can result in contralateral RAPD without visual field defect as the terminal portion of the afferent pupillary pathway is affected [1].

In this case, the lesion is localized at the contralateral middle cranial fossa which is large enough to cause displacement of the midbrain away from the midline. However, factors of increased CSF pressure affecting more on the contralateral optic nerve should be taken into consideration to explain the presence of a contralateral RAPD in this case, although the visual field demonstrated incongruous left homonymous hemianopia. There was no bowtie optic atrophy seen in eye with RAPD, possibly because of the early presentation, allowing near complete resolution of all ophthalmic features in this patient. Younger age, early presentation and

early surgical excision may yield better visual outcome and optic disc changes before optic atrophy sets in [5]. These are among good prognostic factors apart from tumour size, tumour location and extension, preoperative visual status, duration of symptoms, and the surgical technique [6].

## CONCLUSION

Contralateral optic neuropathy may be seen in a tumour causing bilateral disc swelling. Early treatment and excision of a space-occupying lesion may result in better visual outcome, reversal of visual field defect and RAPD.

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