

Atypical presentation of suprasellar meningioma

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ABSTRACT

Demyelinating optic neuritis is the commonest cause of optic neuropathy in young adults. Retrobulbar optic neuritis is one of the frequent clinical forms of demyelinating optic neuritis. We report a case of a 28-year-old lady who presented with blurring of vision in the right eye for one month and pain on eye movement. Fundi examinations were normal. Initial suspicion was that of retrobulbar optic neuritis. Interestingly, imaging showed a suprasellar mass consistent with a meningioma. The patient underwent surgery but unfortunately the excision was incomplete due to a portion of the tumour adherent to the internal carotid artery. Histology confirmed the diagnosis of suprasellar meningioma. The patient was followed up for one year post-surgery with no visual improvement. This case highlights that although rare, suprasellar meningioma should be included in the differential diagnosis of retrobulbar neuritis in young patients.

Keywords: Demyelination, intracranial neoplasm, meningioma, optic neuritis, retrobulbar

INTRODUCTION

Meningiomas are slow growing, mostly benign, tumours that arise from the dura mater. They account for 12 to 35% of all intracranial tumours.¹⁻⁵ Half of these are situated in the anterior half of the brain (usually at the base).¹ Although a study in the United States showed that the prevalence of meningioma was higher in Africans and Caucasians as compared to residents of Asian origin,⁵ Das et al.⁶ showed a much higher prevalence of meningiomas among Asians in Singapore as compared to the Western population. The mean age of presentation among Asians is

63.2 years in males and 53.6 years in females.⁶ The commonest location of meningiomas is parasagittal (25%). Suprasellar meningiomas account for only 9% of all meningiomas.⁷

Demyelinating optic neuritis is the commonest cause of optic neuropathy in young adults. Retrobulbar optic neuritis is one of the frequent clinical forms of demyelinating optic neuritis.⁸

We report the case of a 28-year-old Malay lady who presented with symptoms of retrobulbar neuropathy that was initially suspected to be retrobulbar neuritis in one eye and a visual field defect in the other eye. Investigations later revealed that she had

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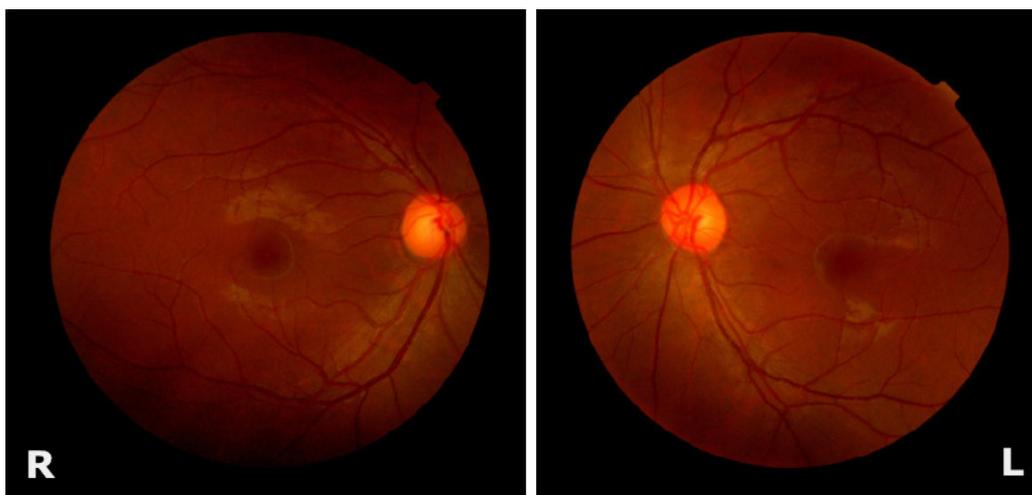


Fig. 1: Coloured fundus photograph showing a normal-looking optic disc in both eyes.

suprasellar meningioma.

CASE REPORT

A 28-year-old female patient walked in to Eye clinic in RIPAS hospital, Brunei Darussalam with blurring of vision in the right eye for one month. The deterioration of vision was slowly progressing in the first three weeks, but rapidly deteriorated in the last week. She also complained of headache and eye pain, related to eye movement. Otherwise, she had no significant ocular or medical history.

On examination, visual acuity was 'Counting Fingers' at two metres in the right eye, and '6/6' in the left eye. Extraocular movements were normal in both eyes with no evidence of proptosis. Anterior segments were normal in both eyes. Intraocular pressure was 16 mmHg in the right eye and 18 mmHg in the left eye (normal intraocular pressure ranges from 8mmHg to 20mmHg). Pupils were round in both eyes and showed relative afferent pupillary defect (RAPD) in the right eye. Fundus examination showed normal-looking optic discs and retinae in both eyes (Figure 1). Cranial nerves examination

was normal on both sides, and systemic neurological examination was also normal.

Colour vision was checked separately for each eye using Ishihara test. It was found to be defective in the right eye, and normal in the left. Visual field examination of the right eye was not possible due to the poor vision. The visual field of the left eye on the other hand, showed visual defect involving the supero-temporal quadrant (Figure 2).

She was admitted to the eye ward. Full blood count, random blood glucose, blood urea and serum electrolytes were taken on admission to rule out possibility of infectious and/or diabetic aetiology, and found to be normal. In view of the young age, female gender and eye pain on movement, intravenous Methyl Prednisolone (1gm daily for three days) was started. A computed tomography (CT) scan of brain and orbits were done on the same day of admission to rule out the possibility of a space occupying lesion. The CT scan showed a mass in the suprasellar area (2.01 cm in diameter) with sellar extension, producing pressure effect on the optic

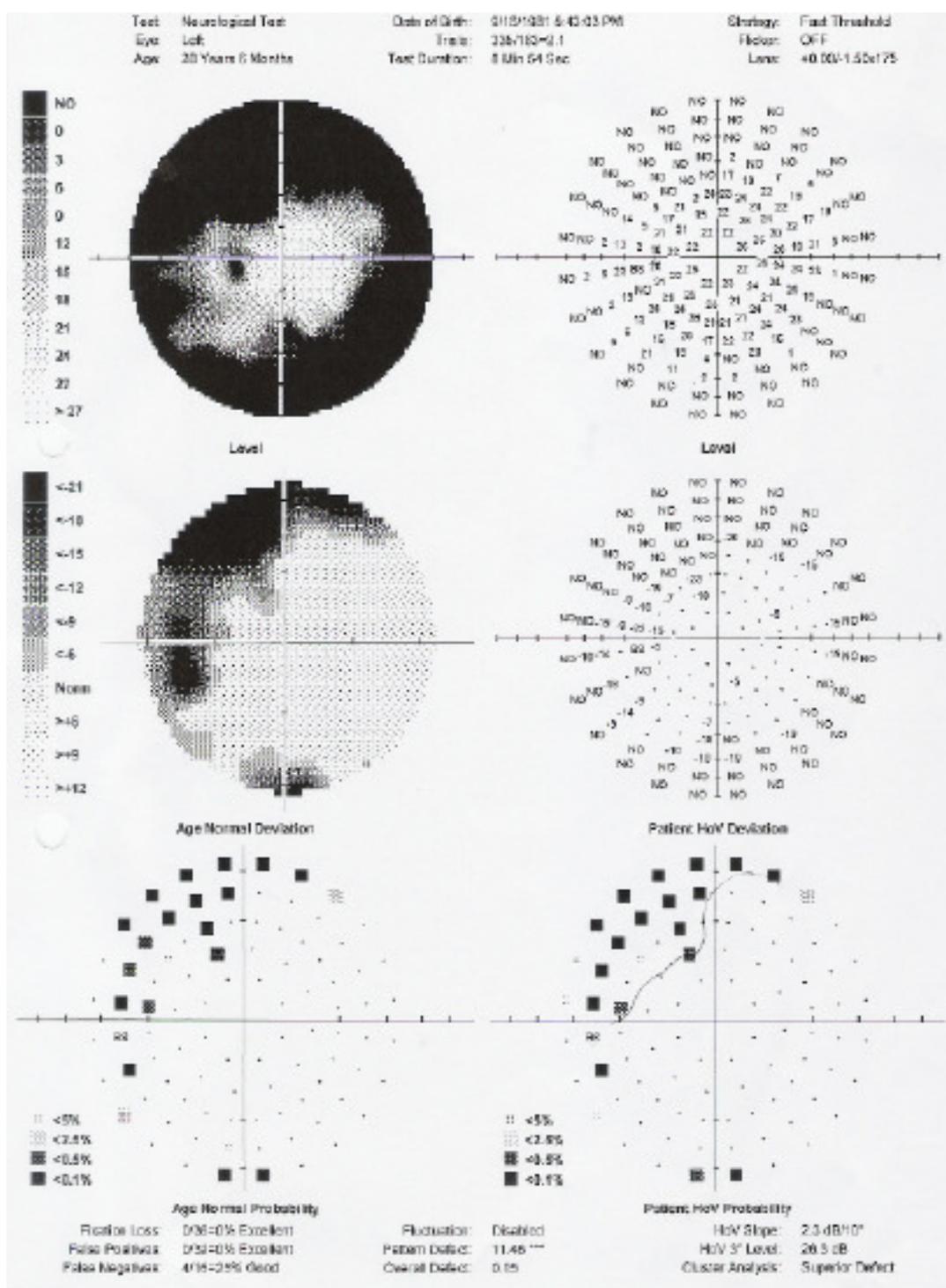


Fig. 2: Automated visual field analysis of the left eye showing a superotemporal quadrantanopia. (Refer to supplementary text for example of other visual field defects).

chiasma (Figure 3). Intravenous methylprednisolone was then stopped after the first dose, and she was referred to the Neurosurgeon for joint management. A Magnetic Resonance Imaging (MRI) was done the

following day, and it confirmed the presence of the suprasellar mass, suggesting suprasellar meningioma (Figure 3a and b).

A pterional craniotomy was done two

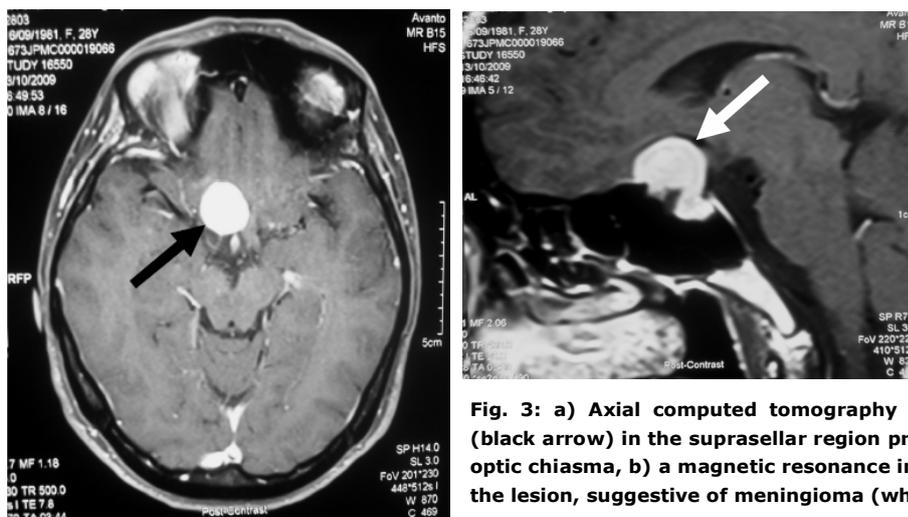


Fig. 3: a) Axial computed tomography image showing a mass (black arrow) in the suprasellar region producing pressure on the optic chiasma, b) a magnetic resonance imaging (MRI) showing a the lesion, suggestive of meningioma (white arrow).

days later. A near total excision of the mass was done. Unfortunately, the resection was incomplete as a small portion of the tumour was adherent to the internal carotid artery. Histopathology analyses revealed the presence of small round cells with eosinophilic cytoplasm and ill-defined cell borders, confirming the MRI diagnosis of a meningioma (Figure 4).

The patient regained complete consciousness two days after surgery. Postoperatively, her visual acuity remained the same in the right eye, and was found to be 6/9 in the left eye. The right optic disc remained pale, with RAPD in the right eye, and visual field defects remained the same in the left eye. Ocular examination was normal otherwise.

She was discharged 10 days after the surgery. A follow-up CT scan showed no radiological evidence of residual mass. Afterwards, she was regularly followed up in both the Eye and Neurosurgical clinics for one year. Her condition remained stable all through.

DISCUSSION

Optic neuropathy is an unusual presentation in

young adults. Generally, demyelinating optic neuritis is the most common cause in this age group.⁸ However other possible causes of optic neuropathy (such as mechanical compression, ischaemic neuropathy, connective tissue diseases and infectious causes) need to be ruled out before ending up with this diagnosis. In our case, although the clinical picture was consistent with retrobulbar optic neuritis suspicious of multiple sclerosis, the underlying aetiology turned to be compressive optic

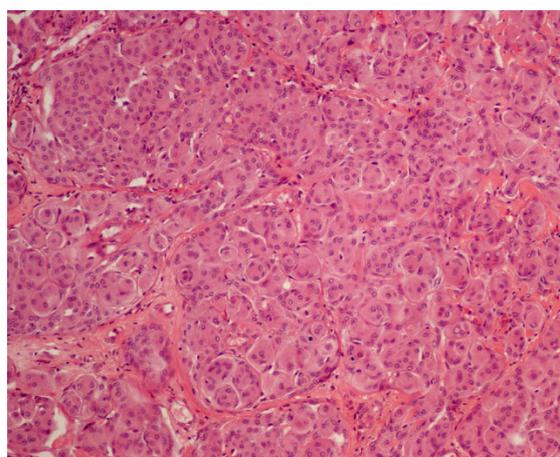


Fig. 4: Histopathological appearance of tumour tissue showing small round cells with eosinophilic cytoplasm and ill defined cell borders, confirming the diagnosis of suprasellar meningioma (H&E stain, x10).

neuropathy secondary to a suprasellar meningioma.

Intracranial meningiomas are the most common primary tumours of the brain, accounting for 33.8% of all brain and central nervous system primary tumours in the United States.¹⁰ In Singapore, meningiomas account for 35.2% of all tumours of the central nervous system.⁶ The population-based prevalence is estimated to be 2.3/100,000 population.¹⁰ In Thailand, meningiomas are found to be the second commonest suprasellar tumours.¹¹ They are twice as common in females than males.⁶ Typically, the prevalence peaks between the fourth and sixth decades of life.

A suprasellar tumour typically presents with visual loss including field defects (86%), eye pain (9%), diplopia (4%) and/or ptosis (1%).¹¹ Our patient presented with blurring of vision, relative afferent pupillary defect (RAPD), ocular pain on eye movement in one eye and non-specific superior visual field defects in the other eye. The former features, with a normal fundus, suggested the possibility retrobulbar optic neuritis as the most likely diagnosis. However, further investigation revealed the presence of a suprasellar mass.

Suprasellar masses can be neoplastic, vascular, congenital or infectious/inflammatory in origin.¹² The clinical features are variable, and depend on the location and the size of the mass. In general, symptoms and signs can be caused either by neural or endocrinological effect of the mass. MRI is the gold-standard diagnostic tool. However, a CT scan may provide complementary information.¹² In

our case, MRI was not initially done as it was not available in our centre. This showed the possibility of a meningioma. The diagnosis was confirmed after the surgical excision.

The management of a suprasellar mass depends on the nature, size and location. In general, surgical excision should be done if possible. In cases of malignant metastases with a known source, surgical excision may not be required. Radiotherapy and/or chemotherapy may be needed depending on the nature of the mass.¹³ Care has to be taken during surgical excision to avoid trauma to the optic nerve, optic chiasma and carotid arteries. In our patient, we were not able to completely remove the tumour as it was adherent to the carotid artery. Attempted complete resection was associated with risk of injuring the artery. The residual mass was followed up by CT scan over a period of one year, and showed no evidence of progression or enlargement. As the tumour was benign in nature, further management with radiotherapy and/or chemotherapy was not considered.

Although multiple sclerosis is rare among Asians compared to Caucasians, visual loss due to optic neuritis on the other hand is more common in Asians.¹⁴ Hence, in the Asian population, multiple sclerosis should be put in mind when managing young females with acute vision loss.

Following meningioma excision, there may be associated disabilities that involve the eye, neurological and/or endocrinological systems depending on the location of the tumour and its extent. Therefore, patients should be managed and followed up with a multidisciplinary approach. In general, follow up should be

continued for longer periods, since recurrence, although rare can occur over a slow course of time.

The prognosis of a completely excised benign meningioma is excellent.¹⁶ However, a partially excised, multiple and/or malignant meningiomas carry high risks of recurrence. Visual improvement has been reported in 67% following resection.¹⁷ In our case, the presentation was late, one month after onset of the symptoms and this may explain the poor post-operative visual outcome.

In conclusion, our case highlighted that suprasellar meningioma should also be considered in young patients presenting with features of retrobulbar neuritis suspicious of multiple sclerosis.

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