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Case Report

Bilateral Compressive Optic Neuropathy Secondary to Tuberculum Sella Meningioma in Pregnancy

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Abstract

A 37-year-old primigravida in her second trimester presented with bilateral painless progressive visual loss. Her vision was hand motion in both eyes. Both pupils were dilated with sluggish reaction to light. Both fundus appeared myopic with bilateral optic atrophy. Magnetic resonance imaging (MRI) of the brain revealed a suprasellar mass with optic chiasm compression and bilateral optic nerve atrophy. As the mass has compromised her vision, a semi-emergency craniotomy and excision of tumour was performed. Histopathological examination confirmed the diagnosis of low grade meningothelial meningioma. Both mother and foetus were well after the surgery. However, post-operatively her vision remained poor due to optic nerve atrophy.

Keywords: Chronic visual loss, meningioma, optic atrophy, pregnancy, tuberculum sella

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Introduction

Incidence of intracranial tumour in pregnancy is as low as seven cases in 125,00 pregnancies (1). The first case of intracranial tumour in pregnancy was described in 1898 by Bernard (2). Meningiomas are the second most common intracranial tumour in pregnancy after gliomas (3). Meningiomas tend to grow rapidly during pregnancy due to hormonal factor, haemodynamic changes and physiological enlargement of pituitary gland (4). Visual prognosis depends on the size of tumour, duration of symptoms and degree of involvement of optic nerve prior to surgical intervention (5). Here, we report a case of meningothelial meningioma of tuberculum sella in pregnancy presented with poor vision.

Case Report

A 37-year-old primigravida at 22-week gestation, presented with bilateral, painless progressive loss of vision for 2-months duration. The visual loss was described as generalized and with similar onset in both eyes. She is highly myopic of -10.0 Dioptre bilaterally. She denied seeing any floaters or flashes of light. There was no history of recent ocular trauma. She had no symptoms of increased intracranial pressure such as headache, nausea or vomiting. She denied any episodes of seizures, limb weakness or numbness.

Her visual acuity (VA) was hand movement in both eyes. Relative afferent pupillary defect was present in the left eye. Both pupils were 7mm dilated with sluggish reaction to light. Anterior segment examinations and intraocular pressure (IOP) were normal in both eyes. Fundus examination showed bilateral optic atrophy (Fig. 1). The optic discs showed myopic appearance with tilted optic discs and presence of peripapillary atrophy. Retinae appear normal in both eyes.

Optical coherent tomography (OCT) of retinal nerve fibre layer (RNFL) showed severe thinning of RNFL bilaterally in all quadrants. Magnetic resonance imaging

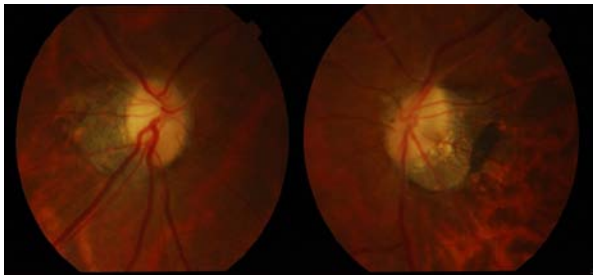

Figure 1: Colour fundus photograph showed with bilateral optic atrophy with peripapillary atrophy

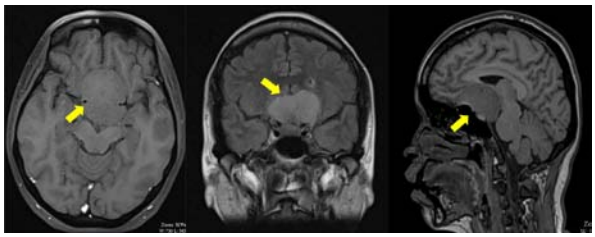

Figure 2: T1 weighted image of MRI brain in axial, coronal and saggital section showed a large suprasellar mass, isointense to the brain (yellow arrow)

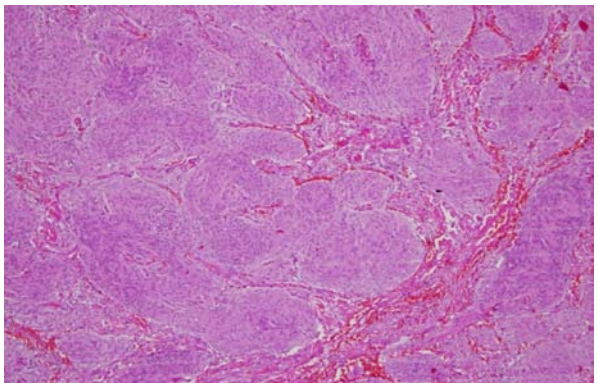

Figure 3: H&E stain (x10) shows whorling and fascicular pattern of the meningioma

(MRI) of the brain showed an intracranial mass with epicentre at suprasellar region, encasing bilateral cavernous portion of the internal carotid artery, and compressing the optic chiasm (Fig. 2). Both optic nerves appeared small in calibre suggestive of optic nerve atrophy.

The diagnosis, treatment options and prognosis of both mother and foetus were discussed among obstetrician, neurosurgeon and anaesthesiologist. Both patient and husband were counselled prior to surgery. She underwent craniotomy and excision of tumour three days after presentation in attempt to salvage her vision. Intraoperatively, the tumour was found to be arising from tuberculum sella extending to retroclival area. The tumour was soft, greyish and highly vascularized. Right optic nerve appeared to be very thinned out, however left optic nerve was preserved. Total resection of tumour was achieved.

Histopathological examination of the tumour showed whorling and fascicular pattern of the tissue (Fig. 3). The tumour cells displayed uniform spindle cells with indistinct cytoplasmic borders forming syncytium in areas with occasional intra-nuclear inclusion seen. Diagnosis of meningothelial meningioma (World Health Organization grade 1) was made. The tumour has low proliferative index of 2% with Ki67.

Post-operatively, both mother and foetus were stable. Unfortunately, her vision remained poor with only hand motion in both eyes due to long standing compression of the optic nerve.

Discussion

Tuberculum sella meningioma is more common in women compared to men. In non-pregnant women of childbearing age, incidences of meningiomas were estimated to be around 1-4.5/100,000 cases (6). Typically, meningioma is a slow growing tumour with insidious onset. However, in pregnancy, meningioma growth may accelerate rapidly (7). The growth of the tumour was thought to be mediated by hormonal changes during pregnancy. Eighty-percent of benign meningioma expresses progesterone receptor (8). The peak of progesterone in second trimester is said to be responsible for the acceleration of tumour growth (8). In our patient, acceleration of tumour growth was evidence by rapid deterioration of visual symptoms.

Haemodynamic adaptation plays a big role in the rate of tumour growth, which also peaks during the second trimester. In second trimester, blood vessels engorge and the plasma volume and red cell mass increases to meet the demand. Perfusion to tissues increases, and so does to the tumour tissues (4). Intra-operatively, the tumour was found to be highly vascularized. Ebner postulated that enlargement of pituitary gland during pregnancy in presence of tuberculum sella meningioma might give additional compression effect that contributes to acceleration of visual loss in the patient (9). Dinc et al in his study showed the pituitary gland volume increased by 120% during pregnancy (10). Despite inability to visualise the configuration of pituitary gland and its infundibulum in the MRI, there is possibility of compression effect by the physiological enlargement of pituitary gland.

A study by Kanaan et al. reported majority patients with meningioma involving tuberculum sella, sphenoid wing, convexity, parasellar, cerebellopontine

angle and anterior falx presented with visual disturbances (11). They might not realize any visual changes at the initial stage until the tumour is large enough to cause compressive symptoms. Chronic compression and late presentation may compromise the visual outcome as most of the optic nerve fibres have demised. In addition, patients might present with symptoms of increase intracranial pressure, such as headache, seizures, nausea and vomiting (3).

Multidisciplinary approach is very important in managing this patient. The goal of the surgery was to salvage patient's vision. Apart from counselling the patient regarding surgery, guarded visual prognosis post-operatively also need to be informed. Besides, emotional support is very important, as she was a primigravida in advanced age. Explanation regarding risk of preterm delivery, and disability to see her own newborn should be addressed to avoid post-partum depression (12).

A study by Palani A found that 27% of patients showed visual improvement post-surgery (13). They found that factors influencing visual improvement post operatively were related to duration of visual symptoms, tumour size, perilesional oedema, brain-tumour interface, arterial encasement and grade of tumour excision (5,13). Zevgaridis et al. described severe pre-operative visual loss appeared to be poor prognostic factor in post-operative visual outcome (14). In our patient, despite poor potential for visual recovery, surgery was performed early to salvage the remaining minimal amount of eye sight that she had to prevent further damage of the nerve fibres due to tumour compression.

Conclusion

This case highlight an example of an aggressive pregnancy accelerated meningioma with significant morbidity. Multidisciplinary approach is important in such case to reduce mortality and morbidity to both mother and foetus. Visual prognosis depends on the degree of involvement of optic nerve. In a patient with poor visual prognosis emotional support is important in view of the risk of postpartum depression.

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