Papilledema with acute profound visual loss in capillary haemangioma of the sphenoid sinus with sellar and parasellar extensions – a triple rarity presentation

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Abstract

Intracranial capillary haemangioma (ICH) is rare. We report a unique case of a young man with ICH who was presented with papilledema and acute profound visual loss. A 27-year-old man presented with bilateral eye reduced vision for 2 weeks with headaches. Examination showed optic disc swelling with severe visual loss. Neuroimaging revealed a vascular tumour epicentre at the sphenoid sinus with sellar and parasellar extensions. The patient developed massive bleeding during transsphenoidal surgery, in which intraarterial embolization was done, followed by a second surgery. Histopathology revealed benign capillary haemangioma. There was minimal visual improvement. He then developed tumour recurrence at 4 months postoperatively. ICH may cause rapid and acute blindness. Management is challenging with a guarded visual prognosis.

Keywords: Intracranial, capillary haemangioma, papilledema

INTRODUCTION

Acute severe visual loss in papilledema is a rare neuro-ophthalmic manifestation. Common diseases associated with bilateral disc swelling accompanied by profound visual loss include severe demyelinating optic neuritis in myelin-oligodendrocyte glycoprotein antibody disease or neuromyelitis optica, toxic, infiltrative, and paraneoplastic optic neuropathy. Generally, patients with papilledema retain normal visual functions. By and large, poor vision will occur when the optic nerve becomes atrophied following its sequelae. Herein, we report a unique case of papilledema with profound visual loss in a young adult as a presenting sign of capillary haemangioma of the sphenoid sinus with sellar and parasellar extensions which then developed as a tumour recurrence.

CASE REPORT

A 27-year-old man presented with two weeks history of progressive blurring of vision which started on the left eye and progressed bilaterally. This was associated with constant, generalised headaches accompanied by tinnitus. Otherwise, there was no pain during eye movement, and no nasal or neurological symptoms, despite worsening vision. Visual acuity was 2/60 on the right and non-perception to light (NPL) on the left. Ocular motility was full. Fundus examination showed optic disc swelling with no macula oedema (Figure 1). Other neurological and systemic examinations were normal.

Magnetic Resonance Imaging (MRI) of the brain and orbit showed a huge lobulated mass with tumoral bleed, epicentre at the sphenoid sinus with sellar and parasellar involvement. CT angiogram demonstrated intrinsic vascularity of the tumour, with peripheral calcification and destructive changes to the surrounding bony structures. Intracanalicular of the optic nerve was compressed by the tumour more towards the left side, causing severe visual loss bilaterally (Figure 2). Full blood count, renal profile, liver function test, pituitary profile and tumour marker...
results were within normal limits.

Endoscopic trans-sphenoidal surgery (ETS) revealed a firm vascularized tumour with blood clots on the surface and within. There was massive bleeding of 8.2 litres intra-operatively, which required stabilization. Later, cerebral angiography and partial embolization of the tumour were performed. Repeated ETS was done after two weeks with tumour debulking, leaving only the capsule as it tightly adhered to the suprasellar region. Histologically, tumour composed of proliferating thin-walled vascular channels with no nuclear atypia. The immunohistochemical stained positive for CD-34 and the pericyte were positive for SMA, which was suggestive of capillary haemangioma (Figure 3).

Our patient had only slight improvement of his vision to 2/60 on the right and counting finger on the left. There was a huge tumour recurrence four months postoperatively, but the patient decided not to proceed with any surgical intervention. On the latest follow-up after one year, his visual acuity was counting finger in the right eye, and hand movement in the left eye with bilateral optic atrophy (Figure 4). He remained neurologically stable without any other neurological deficit.

Figure 1. (A) Fundus photo showing bilateral eye Frisen Grade 2 papilledema, (B) OCT demonstrated bilateral disc retinal nerve fiber layer (RNFL) swelling which is consistent with papilledema.

Figure 2. MR contrasted images show (A) intensely enhancing large lobulated tumour epicentred at sphenoid sinus, anterior tumour extension to posterior ethmoidal air cells and compression to intracanalicular segment of bilateral optic nerves (thin arrow) on axial fat saturationT1-weighted and (B) superior tumoural extension to sellar and suprasellar regions (short arrow) on sagittal T-weighted, (C) Computed tomography angiography(CTA) image shows marked intratumoural vascularity (arrowhead) on sagittal 3D reconstructions.
DISCUSSION

Capillary haemangioma is a benign vascular tumour that commonly appears as a strawberry naevus in infancy. It mainly affects the skin and mucous membranes on the head and neck region. ICH is extremely rare, with only 54 documented cases. It commonly involves the extra-axial; for example, the cavernous sinus. Others include cerebral parenchyma, venous sinuses, fourth ventricles and cerebellum. In adults, it affects females more than males, and the elderly are also susceptible to it.

Several postulations suggest that the origin of angioblasts and trophoblasts, along with a defect in the cytokine regulatory pathways would lead to unregulated angiogenesis of haemangiomas. Pregnancy and hormonal cycle also were also linked to the rapid growth of the ICH.

Clinical signs and symptoms are varied and depend on the location of lesion, ranging from asymptomatic to headache, facial pain, seizure, and dysphasia. Diplopia is the most common visual symptom manifestation. Visual impairment although rare, has also been reported. However,
none of the visual loss was associated with papilledema. In pituitary apoplexy, a patient may develop acute visual loss, but most of the time there will be optic disc pallor due to longstanding tumour compression of the visual pathway. To our knowledge, this is the first case of papilledema with severe visual loss resulting from raised intracranial pressure due to ICH’s rapid growth and tumoral bleed. Compression of the optic nerve by the tumour lead to a rapid decline of the bilateral visual functions.

Our case demonstrated the important role of neuroimaging in the diagnosis of vascular tumours, consistent with other studies. Surgical resection is indicated in symptomatic but has a higher risk of massive bleeding as reflected in our case. Cases with incomplete tumour resection or in those cases involving regions of the brain or skull base where a complete resection cannot be accomplished may benefit from stereotactic radiotherapy. We learnt from this case that preoperative embolization is crucial to minimize the intraoperative bleeding risk. Adequate precaution is required in handling vascular tumour, and the team must be mentally prepared for occurrence of bleeding complications that may need further action, not only by the surgeons but also anaesthesiologist, interventional radiologist and supporting team.

Our patient had a minimal visual improvement after the surgery, and subsequently developed tumour recurrence. Recurrences have previously been reported, usually affecting females. This is the first ever case of ICH recurrence in an adult male.

In conclusion, our case demonstrated that sphenoid sinus ICH has the tendency for rapid growth that may lead to acute visual loss. Management is challenging and requires multi-disciplinary expert involvement.

ACKNOWLEDGEMENTS

The authors would like to thank the Director-General of Ministry of Health Malaysia for his kind permission to publish this article.

DISCLOSURE

Financial support: None

Conflict of interest: None.

Ethics: A full and detailed consent from the patient has been taken.

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