CASE REPORT

Advanced Vestibular Schwannoma: A Case of Optic Disc Oedema without Hydrocephalus

Rita Joana Castro Matos^a, Pedro Nuno Beirão Cardoso Quadrado Gil^a, Joana Margarida Silva Pires^{a,b}, and Nádía Lopes^b

^aDepartment of Ophthalmology, Centro Hospitalar do Baixo, Vouga, Aveiro, Portugal; ^bDepartment of Ophthalmology, Centro Hospitalar Médio Tejo, Tomar, Portugal

ABSTRACT

A wide range of pathologies can cause papilloedema. Vestibular schwannoma is a benign and slow-growing tumour that causes symptoms and findings on ophthalmic examination when the diagnostic is delayed. The authors report a case of a 64-year-old male who presented with bilateral disc oedema secondary to a vestibular schwannoma grade 4. Obstructive hydrocephalus was not evident. The authors suggest that cerebrospinal fluid protein may have a role in the formation of optic disc oedema. A right suboccipital craniectomy was performed to remove the tumour, leading to secondary facial palsy.

ARTICLE HISTORY

Received 31 March 2016
Accepted 23 April 2016

KEYWORDS

Cerebellopontine angle tumour; papilloedema; vestibular schwannoma

Introduction

Vestibular schwannoma (VS) is a benign and slow-growing tumour, derived from Schwann cells of the myelin sheath. They arise in the upper or lower vestibular nerve and rarely in the cochlear branch of the eighth cranial nerve. They account for about 8% of primary intracranial tumours² and 90% of all conditions affecting the cerebellopontine angle.¹

The aetiology of these tumours remains unclear,¹ and they most frequently occurs as a sporadic entity.³ The predominant presenting symptoms are unilateral hearing loss and tinnitus.^{1,2,4} Ophthalmic symptoms are less common and include diplopia, blurred vision, obscurations, and tunnel vision.² Ophthalmic findings may be related to the effect of the expanding mass on adjacent cranial nerves or to increased intracranial pressure (ICP), and include decreased corneal reflex, extraocular muscle abnormalities, nystagmus, lagophthalmos, decreased visual acuity, and papilloedema.² At presentation, optic disc oedema may be present in as many as 8% of patients with vestibular schwannoma (VS).¹ The oedema is usually related to intracranial mass effect or compression of the fourth ventricle resulting in obstructive hydrocephalus.

These benign tumours are classified into four grades according to their extension, from a mass exclusively intracanalicular (grade 1) to the most advanced tumour that extends into the posterior fossa and compresses the brainstem (grade 4).¹ Management options include watchful observation with serial scanning, radiosurgery, or surgery.⁴

We report an unusual case of large VS presenting with optic disc oedema with no evidence of obstructive hydrocephalus.

Case report

A 64-year-old Caucasian male with irrelevant past medical history presented to the emergency room complaining of floaters in the left eye. No other ocular symptoms were referred, including diplopia or visual deterioration. He had no nausea, vomiting, or incontinence and complained of no memory problems.

On ophthalmologic examination, best-corrected visual acuity (BCVA) was 20/20 in both eyes. The movements of the eyeballs, pupillary reflex, intraocular pressure, and anterior segment examinations were normal. Fundus examination disclosed severe bilateral disc oedema and retinal haemorrhages (Figure 1). Systemic blood pressure measurement

Figure 1. Fundus photography at presentation, demonstrating in the right eye (a) optic disc swelling, mild vascular tortuosity, and a small dot-like retinal haemorrhage; in the left eye (b) optic disc swelling, mild vascular tortuosity, and flame-shaped retinal haemorrhages.

Figure 2. Brain magnetic resonance images showing an expansive mass lesion of 30 × 24 × 22 mm. The tumour extended into the posterior fossa, compressing the brainstem and shifting it from the midline (tumour grade 4).

was 180/90 mm Hg and neurologic examination was normal. Brain magnetic resonance imaging (MRI) revealed an expansive mass lesion at the right cerebellopontine angle (**Figure 2**). The lesion occupied the right internal auditory channel and caused deformation of the middle cerebellar peduncle. Although the fourth ventricle was partially compressed by the mass effect, obstructive hydrocephalus was not evident. The audiometry test documented hearing impairment, with profound hearing loss involving both conductive and cochlear components on the right side. Hearing in the left ear was normal.

Three weeks after the diagnosis, the patient underwent neurosurgery for mass removal (a right suboccipital craniectomy was performed). Postoperatively, the patient had a facial nerve palsy (grade III according to House-Brackmann classification). After 10 months of follow-up, there were no clinical or

imagiological signs of recurrence or residual tumour. Best-corrected visual acuity was 20/20 in both eyes. Fundus examination revealed no visible retinal haemorrhages following marked regression of papilloedema.

Discussion

Vestibular schwannoma is a benign tumour, which usually presents with unilateral hearing loss.^{1,2,4} Actually, our patient was found to have unilateral hearing loss with years of evolution.

Accurate diagnosis and rapid surgical intervention are necessary for a good prognosis in patients who present with tumours large enough to cause symptoms and findings on ophthalmic examination. Brain MRI is undoubtedly the imaging modality of choice when cerebellopontine angle disease is suspected.

When performed with gadolinium contrast it has nearly 100% reliability, even for small tumours.¹

Optic disc oedema is an unusual presenting feature of VS. It usually indicates increased intracranial pressure (ICP) as a result of obstructive hydrocephalus or a sizeable mass lesion. ICP can arise as a consequence of intracranial mass lesions, disorders of cerebrospinal fluid (CSF) circulation, obstruction to major venous sinuses, and more diffuse intracranial pathological processes.⁵ Cerebellopontine angle tumours may increase ICP in several ways. They can block ventricular outflow from the fourth ventricle, resulting in hydrocephalus. They also can produce problems with CSF absorption from protein leaking into the CSF.^{6,7}

In the present case, disc oedema was present in the absence of obstructive hydrocephalus. A strong possibility is the role of hyperproteinorrhachia in causing CSF stasis. Increased cisterna magna CSF protein levels due to the sloughing of proteins from the large tumour into CSF may probably have caused raised ICP. Unfortunately, we did not carry out an examination of CSF proteins. However, we believe that more emphasis should be given to the role of hyperproteinorrhachia in causing both CSF stasis and in influencing the visual outcome in large VS.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References

- [1] Bento RF, Pinna MH, Neto RV. Vestibular schwannoma: 825 cases from a 25-year experience. *Int Arch Otorhinolaryngol* 2012;16:466–475.
- [2] Van Meter WS, Younge BR, Harner SG. Ophthalmic manifestations of acoustic neurinoma. *Ophthalmology* 1983;90:917–922.
- [3] Harada T, Sawamura Y, Ohashi T, Harada C, Shinmei Y, Yoshida K, Matsuda H. Severe optic disc edema without hydrocephalus in neurofibromatosis 2. *Jpn J Ophthalmol* 1998;42:381–384.
- [4] Virk JS, Tripathi S, Randhawa PS, Kwasa EA, Mendoza ND, Harcourt J. Tumour resection volumes and facial nerve outcomes for vestibular schwannomas. *Indian J Otolaryngol Head Neck Surg* 2014;66:191–195.
- [5] Dunn TL. Raised intracranial pressure. *J Neurol Neurosurg Psychiatry* 2002;73(SupplI): i23–i27.
- [6] Kumar B, Behari S, Jaiswal KA, Jha KV, Bettaswamy G, Singh U. Large and giant vestibular schwannomas: does cisterna magna hyperproteinorrhachia influence visual status? *Acta Neurochir* 2013;155:63–69.
- [7] Grainger J, Dias SP. Case report: optic disc edema without hydrocephalus in acoustic neuroma. *Skull Base* 2005;15:83–86.