



A CASE REPORT OF BILATERAL DISC EDEMA PRESENTING AS THE FIRST MANIFESTATION OF ACOUSTIC NEUROMA.

Ophthalmology

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ABSTRACT

Purpose : To report a case of bilateral disc edema as first manifestation of acoustic neuroma. **Methods:** A Retrospective Case Report. **Result :** A 35 year old female with 4-month history of blurred vision, affecting both eye (BE). Patient also complained of intermittent headache and difficulty in hearing in the right ear associated with tinnitus and dizziness since one year. On examination, her best corrected visual acuity (BCVA) was 20/40 in BE. The pupillary responses to light and near stimulus and colour vision in BE were normal. Fundoscopy revealed gross bilateral optic disc edema (Grade 4). Visual field testing demonstrated enlarged blind spots. The right corneal reflex was absent. Sensation in the distribution of the right ophthalmic and maxillary divisions of the trigeminal nerve was reduced to light touch and facial nerve function was reduced. Sensorineural deafness in the right ear was confirmed by pure tone audiometry. She had no cerebellar signs with intact gag reflex. MRI showed a heterogenous mass lesion at the right cerebellopontine angle, measuring 4.5 X 4.7 X 5cm. The imaging appearance and the clinical picture suggested the diagnosis of an acoustic neuroma causing raised intracranial pressure and the patient was referred for immediate neurosurgical management. To prevent further visual deterioration, posterior fossa craniectomy and excision of the mass was done. **Conclusions:** Most often, the initial symptoms are neurological, including progressive unilateral sensorineural hearing loss, tinnitus, and vertigo as the first complaints. Visual disturbance and optic disc edema are unusual presenting features of acoustic neuromas. In this case, we demonstrate that acoustic neuroma can present with ophthalmic manifestations as presenting complaints like blurring of vision and disc edema.

KEYWORDS

Acoustic neuroma, disc edema .

INTRODUCTION

Acoustic neuroma originate in the eighth nerve sheath just within the internal auditory canal. With time, they extend into the posterior fossa to occupy the angle between the cerebellum and the pons, cerebello-pontine angle. Most cases are unilateral lesions in patients 20 to 60 years of age with a peak prevalence at the age of 40 to 50 years. Acoustic neuromas typically present with slow-progressive loss of hearing and tinnitus. Dizziness may coincide, but true vertigo is an unusual early symptom. Other presenting manifestations of an acoustic neuroma may be depression or absence of corneal sensitivity and numbness of the face due to involvement of the trigeminal nerve, incomplete closure of the eyelid owing to facial nerve dysfunction, diplopia as a result of sixth nerve paresis and clumsiness of the homolateral limbs from cerebellar compression. When the tumour extends medially and starts to distort the brain stem and cerebellum, more dramatic symptoms such as attacks of true vertigo, ataxia with a tendency to fall to the side of the lesion and spastic paresis are produced. If the patient is untreated, further distortion of the brain stem may block the cerebral aqueduct leading to hydrocephalus, headache and papilloedema. However, depending on the direction of tumour development, the order in which these symptoms occur may vary. Expansion from the internal acoustic meatus in an anterior direction will cause early involvement of the trigeminal and abducens nerves. If the tumour grows primarily in a posterior-inferior direction, the glossopharyngeal, vagus and accessory nerves will be affected early. A mid-cerebellar pontine angle development will cause involvement of the vestibular, cochlear, facial and intermediate nerves the anatomical relationship of the eighth nerve to the other cranial nerves. Symptoms associated with acoustic neuroma only rarely prompt the patient to seek an ophthalmic opinion. However, this patient presented in the ophthalmic out-patients clinic with headaches, papilloedema and life-threatening dilated ventricles which required urgent neurosurgical intervention.

CASE REPORT

A 35 year old female had a 4 month history of diminution of vision which was gradually increasing in nature. She also complained of headache intermittently. On further questioning she admitted that a year earlier she had decreased hearing in her right ear associated with tinnitus. She also experienced several episodes of dizziness and described symptoms of vertigo in past. However, these symptoms also

had improved by the time she was examined. She had no nausea, vomiting, or incontinence and complained of no memory loss. On examination she was alert and oriented. Her BCVA was 6/9 bilaterally. Pupillary responses to light and near stimulus, colour vision were normal in both eyes. On fundoscopy, gross bilateral optic disc edema was noted (figure 1).

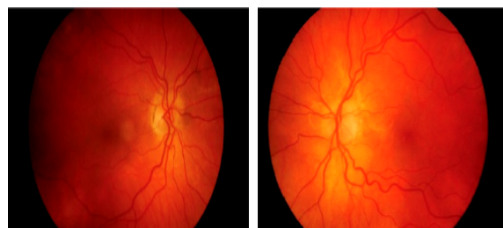


Fig 1 : Fundus photography of BE at the initial visit. Right eye; clear media with optic nerve head edema 360 degree with indistinct margins, mild vessel tortuosity and mild macular mottling. Left eye; indistinct margin superior and inferior of optic disc, indicating disc edema. Retinal vascular is normal calibre and appearance. Macula is evenly pigmented.

Her visual field testing demonstrated enlarged blind spots. The right corneal reflex was absent. Sensation in the distribution of the right ophthalmic and maxillary divisions of the trigeminal nerve was reduced to light touch and facial nerve function was reduced. Sensorineural deafness in the right ear was confirmed by pure tone audiometry. Hearing in the left ear was normal. Her gag reflex was intact and she had no cerebellar signs. The urgently obtained computed tomography (CT) brain scan demonstrated a large mass in the right cerebello-pontine angle with widening and destruction of the right internal acoustic meatus with slight displacement of the fourth ventricle and enlargement of the fourth, third and lateral ventricles (figure 2). Magnetic resonance imaging showed a heterogenous mass lesion at the right cerebellopontine angle measuring approximately 4.5 X 4.7 X 5cm (figure 3). Laterally, it was extending into right internal auditory canal causing its widening and encasing the facial-vestibular nerve bundle, reaching up to the vestibule and modiolus. Inferiorly it was also abutting root entry zone of right 9-10th nerve complex. The diagnosis of an acoustic neuroma causing raised intracranial pressure

was made based on clinical picture and imaging and the patient was referred for immediate neurosurgical management.

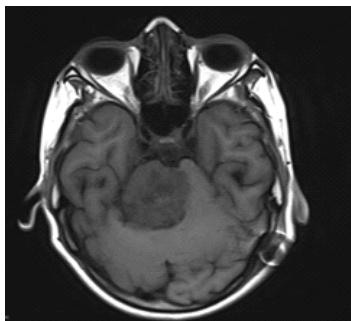


Fig 2 :Axial computerized tomographic scans showing normal ventricular size and displacement of the fourth ventricle

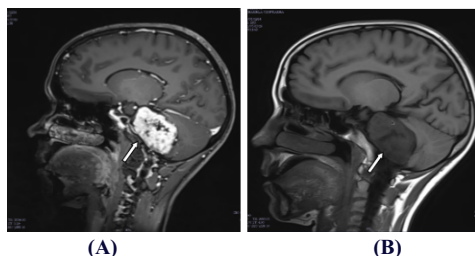


Fig 3 (A & B) :Sagittal T2-weighted magnetic resonance imaging showing a large lesion involving the right side of the cerebellopontine angle and normal ventricular size.

Management options were discussed with the patient. Surgical excision was advised to prevent further visual deterioration. She underwent posterior fossa craniectomy and excision of the mass lesion. Before the lesion was removed, an external ventricular drain and an Intracranial pressure (ICP) monitor were placed. Opening ICP was 14 cm H₂O. Protein concentration of ventricular cerebrospinal fluid (CSF) was 0.47 g/dL. CSF protein concentration from the basal cisterns was significantly raised at 4.07 g/dL.

Patient was operated and gradually weaned off the ventilatory support and was doing well till Post operative day 3. Patient developed high grade fever and started complaining of chest pain and difficulty in breathing and suddenly went into gasping for which she was immediately shifted to Critical Care Unit (CCU). Unfortunately patient went into cardiac arrest and died.

DISCUSSION

Literature shows that about 68% of Acoustic Neuroma patients are diagnosed by otolaryngologists, 28% are diagnosed by neurosurgeons, 19% are diagnosed by neurologists, 5% are diagnosed by physicians, and less than 5% are diagnosed by ophthalmologist, a very low percentage.[1]

A vestibular schwannoma (acoustic neuroma) is the most common tumour of the cerebellopontine angle, corresponding to approximately 8% of intracranial tumours[2,3,4]. A Danish study found an incidence of 200 cases per million inhabitants per year. Most often, the initial symptoms are neurotological, including progressive unilateral sensorineural hearing loss, tinnitus[5,6,7], and vertigo[6] as the first complaints. When it reaches larger dimensions, it can also affect the facial or trigeminal nerve or present central neurological manifestations. Visual disturbance and optic disc edema are unusual presenting features of acoustic neuromas. They usually indicate increased ICP as a result of obstructive hydrocephalus or a sizeable mass lesion.. The core examinations are evoked auditory brainstem response and MRI.

In this case ,we demonstrate that acoustic neuroma can present with ophthalmic manifestations as presenting complains like blurring of vision and disc edema.

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