

Bilateral Optic Disc Edema As Initial Presentation In Case Of Unilateral Vestibular Schwannoma – A Case Report.

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Abstract : We report a case of 55 year old male who presented in ophthalmology outpatient department with complaints of diminution of vision along with headache and giddiness since 20-25 days from presentation. Patient was initially examined in ophthalmology department , and his fundus showed bilateral papilledema. Patient intraocular pressure was within normal limit. Optical coherence tomography(OCT) of patient suggested increased retinal nerve fiber layer thickness. Patient was then send to neurosurgery for second opinion , where he was advised CT scan and MRI with contrast of brain . Later , the scan confirmed the lesion in left cerebello pontine angle with dilated bilateral lateral ventricles and third ventricle suggestive of secondary hydrocephalus. Patient had no complaints of diplopia, it was not associated with facial palsy. Patient complaint of on and off tinnitus.

Vestibular shwannoma is benign and slow growing tumor and ophthalmic manifestation occur in later stage when diagnosis is missed early. Left suboccipital craniotomy and subtotal excision of left vestibular schwannoma was performed and patient was followed up.

Keywords: Bilateral papilledema, retinal nerve fiber layer thickness headache, hydrocephalus, cerebellopontine angle tumor, vestibular schwannoma.

Introduction:

Among cerebello pontine angle tumours vestibular schwannoma is the most common neoplasm and accounts for 8% of intracranial neoplasms⁽¹⁻⁴⁾. Papilledema is uncommon finding in 8% of patients with vestibular schwannoma^(1,5). It is a tumour derived from Schwann cells of myelin sheath. Unilateral variety of vestibular schwannoma are of sporadic origin and bilateral vestibular schwannoma have genetic association and also associated with neurofibromatosis-2. According to extension of tumour mass these are classified into four grades, grade 1 being intracanalicular and when tumour extends to posterior fossa and compresses brain stem it is grade 4⁽⁸⁾. Grade 4 being the most advanced stage of the same.

Initial and predominant presenting symptoms are hearing loss and tinnitus. Ophthalmic manifestations like diplopia, blurring of vision, tunnel vision, restriction of eye movements are rare⁽⁹⁾. When tumour starts compressing brainstem in later stages, it causes raised intracranial tension and thus papilledema⁽⁹⁾.

In this present case, patient initially presented with marked blurring of vision and on findings there was bilateral papilledema and increased retinal nerve fiber layer thickness on OCT. When further diagnosed patient found to have unilateral left sided vestibular schwannoma. Blurring of vision and bilateral papilledema and tinnitus with no other significant complaint as initial presentation makes this case unusual.

Case History

A 55 year old male patient presented to ophthalmology outpatient department for eye examination as patient complaints of diminution of vision and giddiness since 20-25 days. Patient also complained of on and off tinnitus. Patient's symptoms were insidious in onset and symptoms progressed gradually. Presenting symptoms were not associated with diplopia, deviation of angle of mouth, numbness sensation, no restriction of eye movements. There was no history of trauma in past month, no history of hypertension, no history of paralysis. No history of nausea, vomiting, incontinence or loss of memory in past. Patient was not on any medication in past.

On ophthalmic examination:

Right eye:

The best corrected visual acuity (BCVA) was counting finger 3M with improvement till 6/18 with pinhole.

On Slit Lamp Biomicroscopy -

Lid, conjunctiva, cornea, anterior chamber, iris were normal. Papillary was normal size reacting to light. No abnormality was detected in lens. Intraocular pressure when measured at the time of presentation was 12 mm of hg. There was no restriction of extraocular movements.

Fundus examination-

Media was clear

Disc was hyperemic, all margins were blurred, cup disc ratio was obliterated.

Blood vessel- tortuous and dilated vessel with retinal hemorrhage superiorly seen.

Foveal reflex was dull.



Left eye- Best corrected visual acuity was 6/36 and improvement with pinhole was 6/18
On slit lamp examination: lid, conjunctiva, cornea, anterior chamber, iris were normal. Pupil was normal size reacting to light and there was no abnormality detected in lens. Extraocular movements were free and full in all directions of gaze. Intraocular pressure at the time of presentation was 15mm of hg.

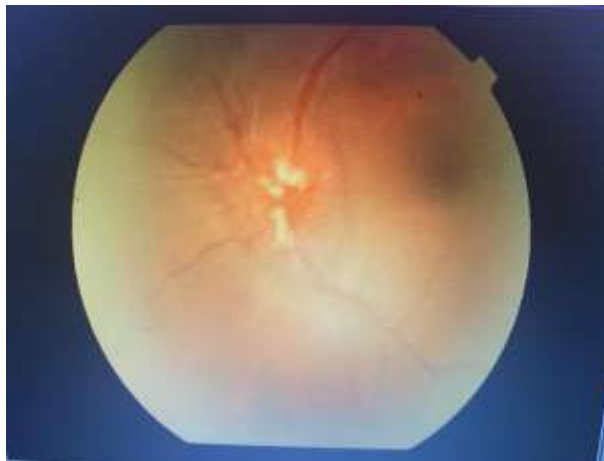
Fundus examination-

Media was clear

Disc was hyperemic , all margins was blurred, cup disc ratio was obliterated.

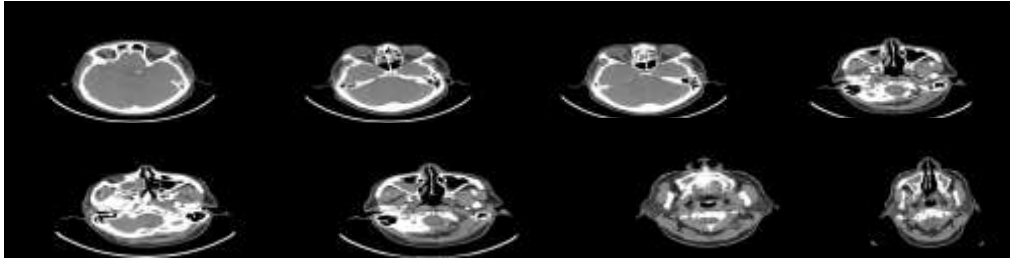
Blood vessel appeared tortuous and dilated.

Foveal reflex was dull.



Patient laboratory investigations were send – complete blood count, liver function test, kidney function test, electrolyte profile, coagulation profile, all were within normal limit. In cerebrospinal fluid glucose was found to be slightly raised, lactic dehydrogenase was below normal limit and protein concentration was normal.

Patient CT brain was done which showed heterogeneously hypodense area at left cerebello-pontine angle with multiple blood density foci and air density foci causing mass effect in the form of compression of fourth ventricle and adjacent cerebellum. These findings were confirmed by MRI brain with Contrast, which suggested extra axial well defined heterogeneously enhancing altered signal intensity mass lesion in left cerebello-pontine angle, extending into porus acousticus causing its widening with presence of mass effect suggestive of vestibular schwannoma. Secondary hydrocephalus with mild periventricular ooze was also observed.



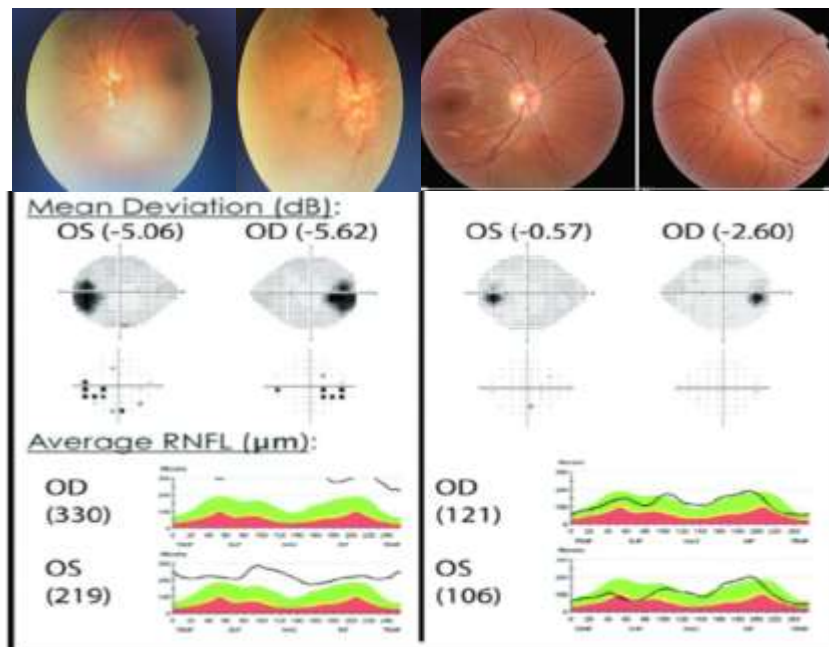
Patient underwent surgical intervention after 2 weeks of diagnosis for the same which was left suboccipital retrosigmoid craniotomy and subtotal excision of left vestibular schwannoma with left lateral ventricle with external ventricular drain was performed. Brownish black tissue was retrieved after surgery which was send for histopathology and reports suggested schwannoma.

Patient was then followed up, there was no evidence of facial palsy after surgery, though patient complaint of difficulty in hearing .

Patient was followed up for one year and gradual improvement in vision to 6/9 which improved to 6/6 with pinhole was observed. On fundus examination retinal haemorrhages dissolved with resolving hyperaemia and disc margins becomes well defined.

Patient optical coherence tomography was done preoperatively and it showed average retinal nerve fibre layer to be 330 μm right eye and 219 μm in left eye.

Patient was followed for 1 year postoperatively and 1 year after resection of tumour optic disc oedema resolves and retinal nerve fibre layer thickness reduced to 121 μm in right eye and 106 μm in left eye.



Discussion:

Bilateral papilledema in young patients suggest early sign of ventricular dilatation. Papilledema is mainly a feature of raised intracranial tension occurs often obstructive hydrocephalus , venous obstruction , pseudotumour cerebri or supratentorial mass effect⁽¹⁾. In this case T2 weighted image in MRI suggestive heterogenously hyperintense mass lesion

which extends to porus acoustic causing widening. Lesion has a mass effect in the form of displacement of adjacent cerebellar folia and fourth ventricle . Thus, explaining the cause of unilateral vestibular schwannoma causing bilateral papilledema.

Optic disc oedema is an unusual finding in patients with vestibular schwannoma. It is generally a feature of raised intra cranial pressure which is directly proportion to size of the lesion and obstructive hydrocephalus⁽⁸⁾. Cerebello pontine tumour causes ventricular obstruction from fourth ventricle resulting in hydrocephalus^(8,9,10). There is protein leakage in CSF hence hindering its absorption, thus increasing more intracranial pressure. Here , in our case patient intraocular pressure was normal in both eyes and at the time of surgery intracranial pressure recorded was normal, this suggests patient could have normal pressure hydrocephalus^(10,11). But, other symptoms related with normal pressure hydrocephalus was absent like gait ataxia, memory loss and incontinence was absent. Chhablani et al reported a rare case of Schwannoma of the Larynx⁽¹²⁾. Proper evaluation of symptoms and signs and detailed examination and investigation leads to early detection⁽¹³⁻¹⁵⁾. Post-operative evaluation and follow-up of cases are equally important⁽¹⁶⁻¹⁸⁾.

Vestibular schwannoma being the benign tumour which progresses slowly , and thus diagnosis of such cases at early stages becomes important as in lateral stages it can progress to pseudotumour cerebri syndromes causing decompensation, hence , resolution of disc oedema even after resection of tumour is not seen.

In this case , early diagnosis of schwannoma and its management provided us with better visual outcome with resolution of disc oedema with decrease in thickness of retinal nerve fiber layer on OCT on follow up.

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