

Acoustic schwannoma: An unusual presentation

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ABSTRACT

Acoustic schwannomas are the benign tumors arising from vestibular division of 8th cranial nerve. The worldwide incidence is 1 in 1,00,000 population. This accounts for about 9% of all intracranial tumors and about 90% of all cerebellopontine (CP) angle tumors. We report a rare case of unilateral acoustic schwannoma with involvement of pupil and with 3rd cranial nerve palsy; but without evidence of any adjacent cranial neuropathies and hydrocephalic changes. The exact mode of association between the tumor and oculomotor nerve involvement could not be established.

Keywords: acoustic schwannoma, vestibular schwannoma, oculomotor nerve palsy

INTRODUCTION

Acoustic Schwannoma is a benign and usually slow-growing tumor that develops from the 8th cranial nerve root. This accounts for about 9% of all intracranial tumours and about 90% of all cerebellopontine (CP) angle tumors. Also known as vestibular schwannoma, it sometimes affects the other cranial nerves. Cordulla and Madjid in their series of 1000 cases of acoustic schwannomas found nerve involvement of acoustic (95%), vestibular (61%) followed by trigeminal (9%) and facial (5%) nerves.¹ Though a slow growing tumour, in few cases it may grow rapidly and become large enough to press against the brain and interfere with vital functions.

The signs and symptoms of acoustic schwannoma manifest from the tumor pressing on the adjacent nerves, nearby blood vessels or brain structures. As the tumor grows, it is more likely to cause signs and symptoms, although tumor size doesn't always determine its effects. It's possible for a small tumor to cause significant signs and symptoms, which may include hearing loss, tinnitus in the affected ear, unsteadiness, loss of balance, dizziness, vertigo, facial numbness and weakness. In rare cases, an acoustic schwannoma may grow large enough to compress the brainstem and becomes life-threatening. We report a rare case of acoustic schwannoma associated with ipsilateral oculomotor nerve involvement.

CASE REPORT

A 34 year male patient, who was asymptomatic until six months ago, noticed difficulty in hearing on right side. He has had the habit of taking bath in pond water. Subsequently he developed discharge and pain in right ear. For these symptoms he consulted an otolaryngologist who diagnosed him to be suffering from small central perforation of tympanic membrane in right ear. Findings in left ear were normal. The condition was medically treated.

After about a month, he again noticed progressive deafness in right ear. The magnitude of symptoms gradually increased over months. Examination revealed that both tympanic membranes were intact though on right side it showed evidence of old healed perforation. Both Rinne's and Weber test pointed towards right sided sensorineural loss. By that time he also developed tinnitus in right ear. About a month later, he developed occasional ataxia with diplopia for which he was referred to a neurologist and an ophthalmologist. There was no clinical evidence suggestive of increased intracranial pressure.

Examination revealed unaided 20/20 vision in both the eyes. On right side there was complete ptosis with no levator function. Right divergent squint measuring more than 40 prism diopter was observed. There was complete paralysis of all the extraocular muscles of the right eye except lateral

rectus and superior oblique. Fundus examination in both the eyes was normal. Pupil on the involved side was dilated but corneal sensation was preserved.

A contrast enhanced CT scan using 5mm x 10 mm section in axial plane showed a mass lesion in cerebellopontine angle on the right side. The fourth ventricle was slightly compressed by an increased attenuating enhancing mass at right CP angle area. The third and fourth ventricles were mildly dilated with septum in the midline. The patient was referred to department of oncosurgery, where he was advised to be on close observation.

Figure 1: CT scan showing the mass lesion



DISCUSSION

Schwannomas are the tumors arising from Schwann cells of nerve roots. Most frequently they involve eighth cranial nerve followed by fifth cranial nerve. They rarely arise from optic and olfactory nerve since they are myelinated by oligodendroglia rather than Schwann cells. Bilateral schwannomas are commonly associated with neurofibromatosis type-2, however, our patient did not reveal any feature suggestive of the same. Acoustic schwannomas typically arises from vestibular division rather than acoustic division of eighth cranial nerve. The most common presenting symptoms are progressive unilateral

sensorineural deafness with tinnitus or both as our case presented with. Among other signs about 70% of patients exhibit some evidence of ataxia.² Our patient developed ataxia about two months after he developed unilateral deafness. Schwannomas are typically slow growing benign intracranial tumours. Though the tumor originates laterally and expands to press on other contents of inner ear namely cochlear and facial nerve. Then the tumor has got the tendency to expand medially into CP angle. At this point of time it comes in contact with fifth and sixth cranial nerve. But downward extension may impinge upon ninth and tenth cranial nerve. Further extension is likely to cause aqueductal compression leading to obstructive hydrocephalus.³ As the tumor grows to impinge upon various adjacent cranial nerves, the patients exhibit clinical features accordingly. Early loss of corneal reflex is classical though was not present in our case. The Schirmer test was also normal in this case. No weakness of facial muscles was observed. No hoarseness was noted neither any nystagmus. He did not report any facial numbness or paresthesia. But, he classically presented with features suggestive of ipsilateral complete third nerve palsy with involvement of pupil. No suggestive history of trauma or systemic hypertension was found. Post-prandial blood sugar level was normal. The reported rates of cranial nerve involvement in case of acoustic schwannoma are acoustic (95%), vestibular (61%), trigeminal (9%) and facial (5%) nerves.¹ Multiple cranial nerves involvement in a case of acoustic schwannoma might present as a false localizing sign.⁴ Literature search failed to reveal any documentation regarding concurrent presentation of acoustic schwannoma with ipsilateral pupil involving oculomotor nerve palsy.

CONCLUSION

Review of standard journals and neurosurgery text books point towards 5th, 6th, 7th, 9th and 10th cranial nerve involvement in cases presenting with acoustic schwannomas. The actual causal association of this tumor with 3rd cranial nerve

involvement could not be properly established. It may be an isolated finding or it may be a false localizing sign.

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