Ancient Schwannoma Originating from Vestibular Nerve: A Case Report

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ABSTRACT

Ancient schwannoma is a pathological subtype of schwannoma. Intracranial ancient schwannoma is a rare benign tumor and there is no detailed report of ancient schwannoma orginating from the vestibular nerve, on reviewing the literature. Such a rare case is reported in this article.

Keywords: Intra Cranial Ancient Schwannoma, Vestibular Schwannoma, Cerebellopontine Angle

*See End Note for complete author details

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INTRODUCTION

Schwannoma, also known as neurilemoma or neurinoma, is a benign nerve sheath tumor arising from differentiated Schwann cells.[1] Ancient schwannoma (AS) is a pathological subtype of schwannoma, first reported by Ackerman and Taylor.[2] Ancient schwannoma (AS) is a subtype of schwannoma characterized by slow progression despite degenerative changes in pathology.[3] It is a benign schwannoma characterized by pathological nuclear atypia or degenerative changes such as hemorrhage with hemosiderin deposition, lymphocytic infiltration, and cyst formation, and paucity of mitotic figures.[2] Here we describe a rare case of an ancient vestibular schwannoma with rapid progression.

CASE REPORT

A 40-year-old female patient presented with complaints of left sided hard of hearing, vertigo, tinnitus, swaying of body to the left side of 6 months duration. She also had occipital headache since 2 months back, and deviation of angle of mouth to right side and difficulty in left eye closure since 2 weeks back. She had been undergoing treatment for hypothyroidism for 4 years. Ear examination demonstrated a normal tympanic membrane and House Brackmann grade 3 facial palsy on left side. Spontaneous nystagmus present with its fast component beating towards right side. Tuning fork test showed positive Rinne's test on both sides, Weber lateralized to right and absolute bone conduction

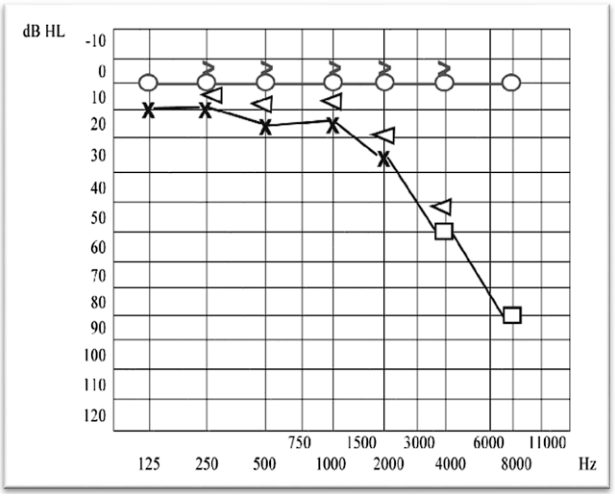

Figure 1. PTA

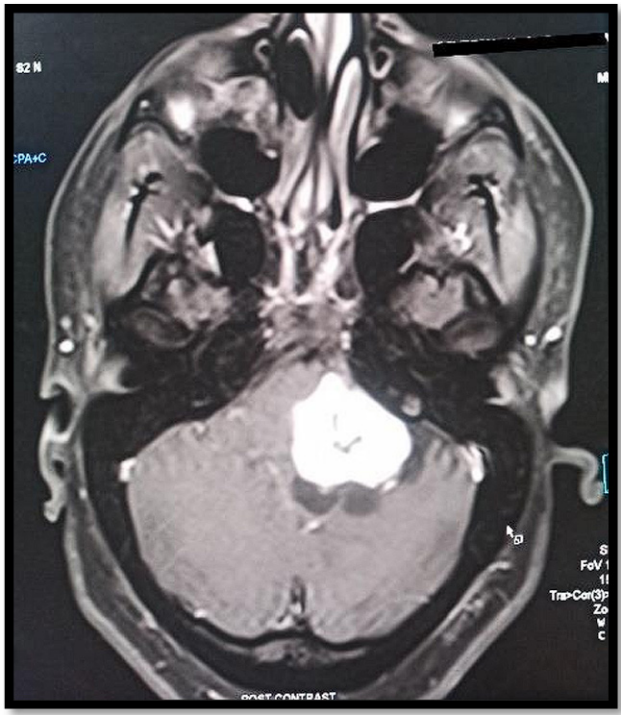

Figure 2. T1 weighted MRI: hypointense lesion

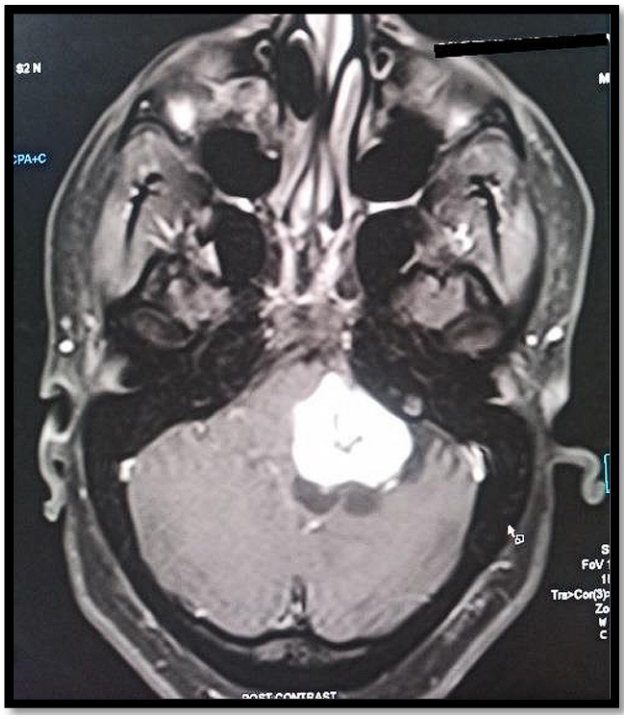

Figure 3. T2 weighted MRI: hyperintense lesion

decreased on both sides. Pure tone audiometry showed down-sloping high frequency sensorineural hearing loss in the left ear **(Figure 1).**

MRI revealed a left cerebellopontine angle (CPA) tumor suspected to be a vestibular schwanomma with extension into left internal acoustic meatus **(Figures 2 and 3),** and mass effect evidenced by buckling of pons, middle cerebellar peduncle, left cerebellar hemisphere and effacement of fourth ventricle **(Figures 4 and 5).**

We resected the left CPA tumor through left retrosigmoid, retromastoid suboccipital approach **(Figure 6)** with intraoperative facial nerve monitoring. Postoperative period was uneventful.

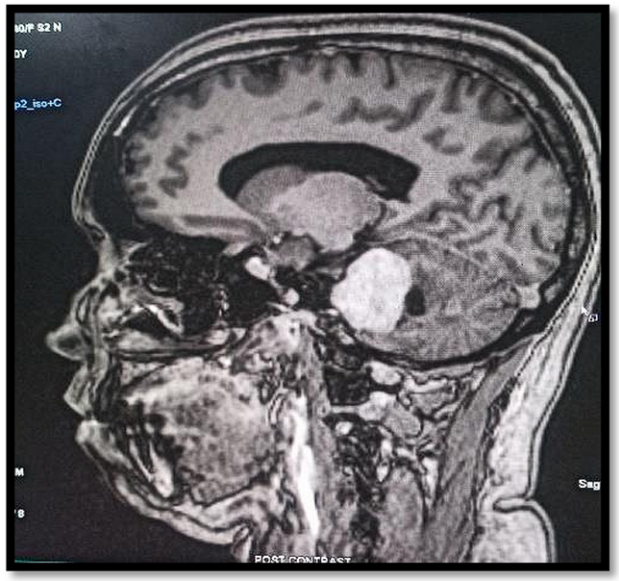

Figure 4. MRI sagittal section

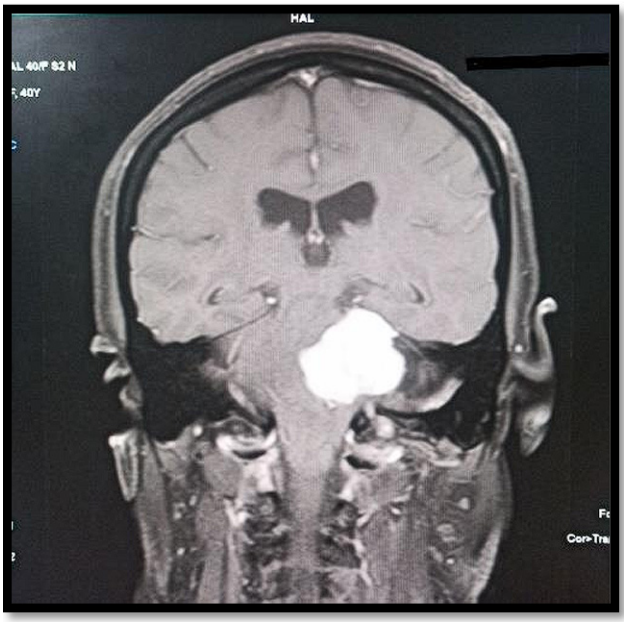

Figure 5. MRI coronal section

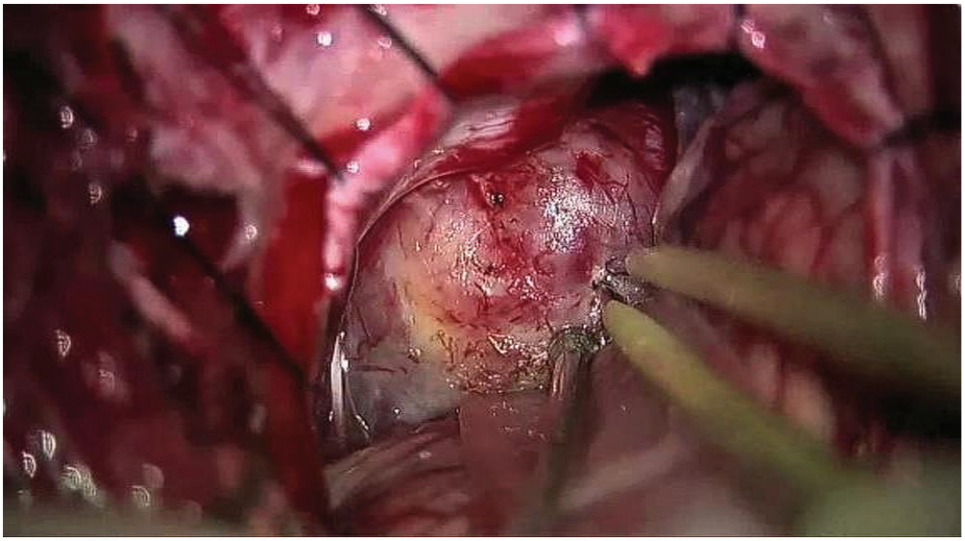

Figure 6. Intraoperative image of left CP angle tumor

Histopathologically, there were hypercellular areas with nuclear palisading and myxoid hypocellular areas **(figure7a)**. There was nuclear pleomorphism and hyperchromatism without mitosis **(figure 7b).** Degenerative changes such as foci of calcification, hemorrhages, cyst formation and hyalinization **(figure 7c).**

Immunohistochemistry positive for desmin **(figure 7d),** S-100 **(figure 7e), SMA (figure 7f).** The tumor was diagnosed as Ancient Vestibular Schwannoma.

DISCUSSION

In 1951, Ackerman et al described a new variant of schwannoma, which showed features of long-standing duration of the tumor in the nerve. Histopathologically vestibular schwannoma showed intermingling of Antony A cellular areas and Antony B cystic areas and Verocay bodies.[4] In addition to this Ackerman noticed grossly larger tumor with microscopic features like, fibrous nodules, hyalinization and fatty degeneration,

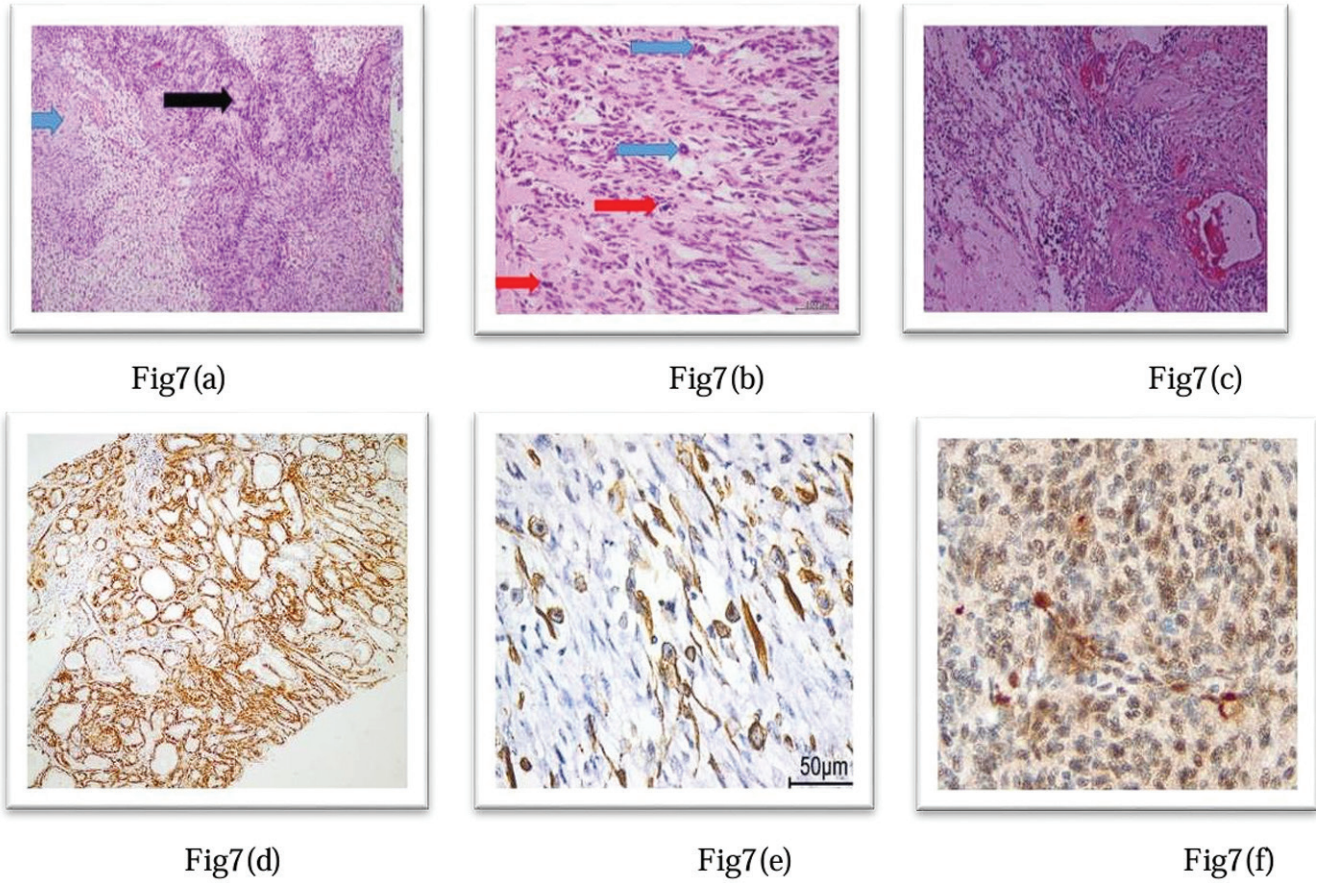

Figure 7. Histopathological features of ancient vestibular schwannoma

which is due to features of long-standing duration like diffuse overgrowth of the tumor and vascularization. Hence, he described a newer entity called Ancient Schwannoma.[5] The long term progression of the tumor is thought to produce degenerative changes, hence the name Ancient.[3] Though it is benign, it is often misdiagnosed as malignant tumor, because it mimics some features of malignancy like nuclear atypia, necrosis and degenerative changes.[6] Ancient Vestibular schwannoma is difficult to diagnose based on clinical and radiological features alone, hence definitive pathological diagnosis is necessary. Pathological features include, nuclear atypia without mitosis, degenerative changes, hemosiderin deposition, inflammatory cell infiltration, which are all a microscopic finding in our case. Immunostaining is positive for S100, SOX10 in general and ours showed positivity for S100, desmin and SMA which are all suggestive of vestibular schwannoma of ancient type.

According to Isobe et al, Ancient Schwannomas are usually located deep in the head and neck, thorax, retroperitoneum and pelvis and extremities of elderly persons. MRI is the most useful technique for the evaluation of ancient schwannoma. Normal schwannomas show peripheral low intensity signal in T1W imaging and high intensity signal in T2W image.[7] In contrary, ancient schwannomas show well circumscribed complex cystic mass with different enhancement pattern which mimics malignant tumors. Hence a strong suspicion of ancient vestibular schwannoma has to be considered as radiological imaging and histopathological features mimics malignancy.

Extracranial cranial lesions such as retroperitoneal cavity reaches substantial size by the time of diagnosis because of nonrestrictive and expandable properties of retroperitoneal space. In contrast intra cranial space is surrounded by skull and the space in the skull base where most schwannomas occur is s especially narrow therefore intracranial ancient schwannoma often presents with neurological symptoms at an early stage of development leading to prompt diagnosis and treatment.[3]

We report a rare case of rapidly progressing ancient vestibular schwannoma. Ugokwe, et al reported the first case of intracranial AS and only three cases of intracranial AS have been previously reported, all of which were trigeminal in origin.[2] Tsuchiya, et al reported first case of intracranial ancient schwannoma arising from the vestibular nerve. To conclude, intracranial ancient schwannoma is a rare benign tumor, so further study and review of cases are necessary to clarify its clinical features. Even though it is a benign pathology, there are cases of rapid growth and early recurrence. Therefore, prompt diagnosis and strict follow-up is necessary even if adequate removal of the tumor is achieved. In addition to pathological studies, the genetic background of intracranial ancient schwannoma warrants future investigations and hopes to add higher treatment modality.

END NOTES

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Conflicts of Interest: None declared.

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