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Case Report

Ancient Schwannoma affecting the intracranial portion of the trigeminal nerve: A case report

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ABSTRACT

Background: Ancient trigeminal schwannomas are extremely uncommon benign tumors. Such tumors are long-standing, slow growing and may demonstrate seemingly malignant features irrespective of its benign nature. The tumor may involve the trigeminal nerve root, the trigeminal ganglion, or any of its peripheral branches. Its clinical presentation may include trigeminal neuralgia, blurry vision, diplopia, or even seizures. Surgical excision is the mainstay of treatment with definite diagnosis only by histopathology.

Case Description: We described a case of a 35-year-old female presenting with recurrent episodes of generalized seizure and left-sided weakness. Brain imaging showed a right temporal space occupying lesion. Results of histopathology were consistent with trigeminal schwannoma associated with ancient histopathological changes. Complete tumor excision was achieved by a two-stage craniotomy, which led to the patient's condition to dramatically improve.

Conclusion: Ancient trigeminal schwannomas are easily diagnosed through histopathology and result in favorable clinical outcomes after total microscopic surgical excision. A high suspicion index of ancient schwannoma diagnosis should be derived from the patient's presenting clinical picture and the classical findings derived from neuroimaging.

Keywords: Ancient schwannoma, Intracranial tumor, Trigeminal nerve

INTRODUCTION

Schwannomas are nerve sheath tumors which develop from Schwan cells and are usually benign. Trigeminal schwannomas are a rare entity, accounting for 0.8–5% of intracranial schwannomas,[2] in which ancient changes are exceptionally uncommon. Clinically, it presents with facial pain, blurred vision, headache, and seizures.[2,7,13] To the best of our knowledge, this is the third reported case to describe an ancient schwannoma involving the intracranial portion of the trigeminal nerve.

CASE PRESENTATION

History and physical examination

A 35-year-old right-handed, medically free female presented to our neurosurgery outpatient clinic complaining of recurrent generalized seizures, chronic facial pain, diffuse dull headache,

blurry vision, and dizziness. On physical examination, the patient was confused, scored 14 out of 15 on the Glasgow Coma Scale. She demonstrated a spastic gait, hyperreflexia, and hemiparesis exclusively on her left side. In addition, she scored four out five on the Medical Research Council power scale. Moreover, the patient showed facial asymmetry secondary to a jaw-related pathology, positive Hoffman, Babinski, and Barre's signs.

Neuroradiology

Preoperative brain computer tomography (CT) scan demonstrated a cystic lesion at the right middle fossa on base of the skull [Figure 1]. Based on these findings, a provisional diagnosis of chordoma versus chondrosarcoma was made. Brain magnetic resonance imaging (MRI) showed a right temporal extra-axial cystic lesion attached to the skull base, measuring 7 × 6 cm at maximum diameter [Figure 2].

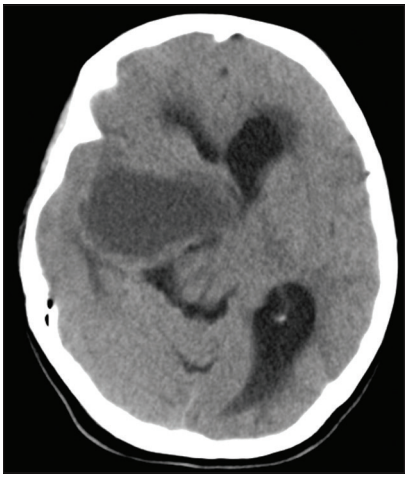

Figure 1: Preoperative noncontrasted brain computer tomography-scan demonstrated a hypodense cystic lesion on the right middle cranial fossa at the base of the skull. The lesion extends upward contributing to a mass effect on the contralateral hemisphere. Notice the contralateral ventricular dilatation.

Management

Patient underwent a two-stage right modified pterional craniotomy, with posterior extension, using a sub-temporal approach. Complete excision was achieved under microscope and navigation guidance. The first stage involved the evacuation of the aforementioned cystic component, which was sent for histopathological examination. Subsequently, a complete tumor excision was achieved in the second stage. Intraoperatively and under microscopic visualization, the third division of the trigeminal nerve was indistinguishable from tumor. Nonetheless, the third division was partially preserved through the operation.

Intraoperative neurophysiologic monitoring (IONM) was not utilized due to a myriad of reasons including but not limited to: lack of a standardized IONM procedures,[12] sensitivity to temperature, demanding anesthetic protocols,[5] and its inability to affect extend of resection.[10]

Histopathological examination of the specimen showed a partially encapsulated tumor, composed of bland looking spindle cells that were arranged in hypocellular and hypercellular areas. Nuclear palisading was present in the hypercellular areas. Minimal nuclear atypia was present but no signs of active mitosis were seen. Moreover, thick walled vessels and evidence of previous hemorrhage were evident. The tumor cells were strongly positive for S100 immunostain (both nuclear and cytoplasmic). Both microscopic and immunohistochemial characteristics of the lesion were consistent with ancient schwannoma [Figure 3].

Follow-up

The patient was followed up for 2 years. On her last follow-up, she demonstrated improved symptoms, no power deficit yet complained of the persistence of the aforementioned right-sided mouth deviation. MRI follow-up showed no signs of tumor recurrence [Figure 4].

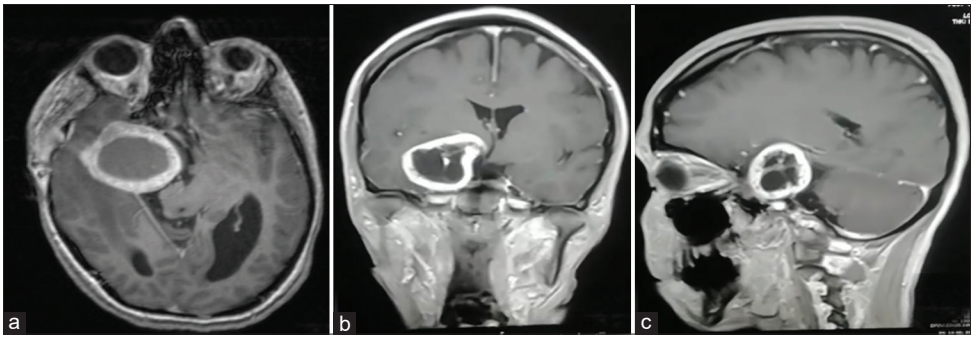

Figure 2: Three views T1 ([a] Axial, [b] Coronal, [c] Sagittal) Contrasted magnetic resonance imaging showing an extra-axial cystic lesion attached to the skull at the right cavernous sinus associated with significant mass-effect, compression of the brain parenchyma, vasogenic edema in the right temporal and parietal lobes, ventricular compression and subfalcine herniation with active hydrocephalus particularly in the left lateral ventricle. The tumor exhibits a close relation to the right Meckel's cave and compresses it.

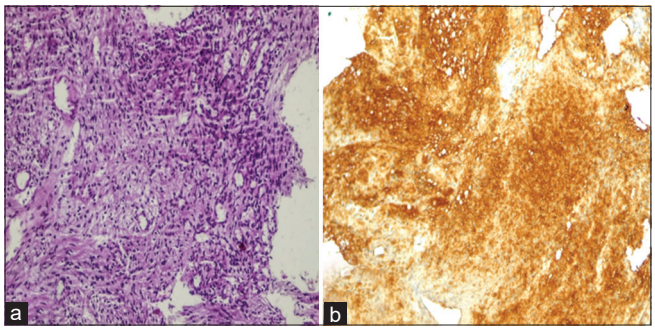

Figure 3: Histopathological findings. (a) Hematoxylin and Eosin stain, showing spindle cells arranged in fascicles in hypercellular and hypocellular areas, with mild nuclear atypia without any signs of active mitosis. Thin-walled blood vessels were noted. (b) Immunohistochemical staining revealed diffuse positive reactions of cells toward the presence of the S100 protein.

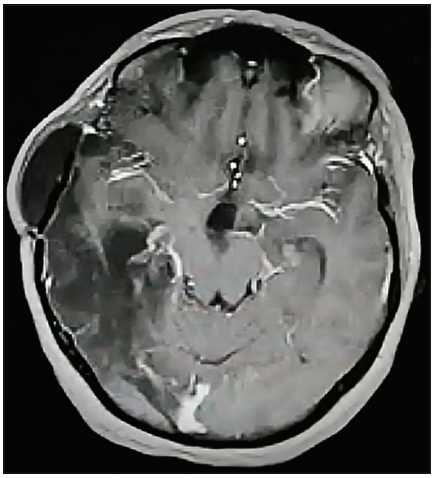

Figure 4: Postoperative axial, T1, contrasted magnetic resonance imaging illustrated postoperative leukomalacia, no residual tumor and the absence of a mass effect in terms of midline shifts or ventricular compression. Postoperative subgaleal collections are noted at the site of surgery.

DISCUSSION

Schwannomas are slow-growing tumors of the perineural Schwan cells. They may involve any myelinated nerve including peripheral, cranial, or autonomic nerves.[2,11] About 25–40% of all schwannomas occur in the head and neck.[6] Schwannomas account for 8–10 % of all intracranial tumors and occur predominantly during the 2nd–5th decade of life, of which 0.8–8% involve the trigeminal nerve. Trigeminal schwannomas are the second most common intracranial schwannoma following vestibular schwannomas. They account for 0.07–0.3% of all intracranial tumors. Trigeminal nerve root, the trigeminal ganglion, or any of the three peripheral branches may be involved. Depending on the site, size, and surrounding structures, these tumors may present with facial hyperesthesia, facial pain, blurred vision, diplopia, headache, seizure, and weakness in the muscles of mastication.[2,7,13]

Ancient schwannomas are a benign uncommon histological variant of schwannomas, first described by Ackerman and Taylor.[1] These tumors are long-standing, slow-growing with degenerative changes. Typically, these tumors show nuclear atypia, hyper-cellular areas, and regressive changes including calcification, cystic formation, hemorrhage, fibrosis, and hyalinization on histopathology. These features closely resemble malignant tumors, however, the presence of a capsule, hemorrhagic areas, degenerative changes, and absence of mitotic activity, support the benign nature of the tumor.[8] On immunohistochemistry staining, these tumors are positive for the S100 protein, which is an important marker to differentiate benign schwannomas from other high grade or malignant lesions.[6] Ancient schwannomas involving the intracranial part of the trigeminal nerve are extremely rare and were reported in the literature only twice.[3,11]

Neuroimaging is of paramount importance in establishing the diagnosis, assessing the involvement of adjacent structures, and to determine the best surgical approach. MRI is the imaging modality of choice, in which T1-weighted imaging of trigeminal schwannomas usually shows low signal intensity, while T2-weighted images show high signal intensity. These tumors exhibit momentous enhancement following contrast injection. Brain CT-scan is particularly important for tumors located near the skull base, as these tumors appear as iso- to slightly hypo-dense lesions with variable enhancement postcontrast injection.[2,4,13]

Complete tumor resection with nerve function preservation, minimal postoperative morbidity and mortality are the present goals of trigeminal schwannoma management. With the advances in microsurgical techniques and skull-base approaches, outcomes have dramatically improved with higher survival rates and lower recurrence rates. However, complete tumor resection could be associated with a variety of complications including meningitis, fistula formation, trigeminal neuralgia, and masseter muscle atrophy.[9,13]

CONCLUSION

Ancient trigeminal schwannomas are benign intracranial tumors. Diagnosis can be made by histopathology without difficulty. Total microscopic resection can be achieved, resulting in favorable outcomes. Ambiguous cases of ancient trigeminal schwannoma could be handled through fastidious analysis of the presenting patient's clinical picture and meticulous evaluation of neuroimaging for classical signs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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