Case Report

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

Abdulrahman Al-Shudifat1, Baraa Mafrachi2, Abdallah Al-Ani2, Amer Khalil Al Qaisi1, Nadwa Bustami3

1Department of Neurosurgery, The University of Jordan, 2Department of Special Surgery, School of Medicine, The University of Jordan, 3Department of Pathology, Microbiology and Forensic Medicine, The University of Jordan, Amman, Jordan.

E-mail: *Abdulrahman Al-Shudifat - a.shudifat@ju.edu.jo; Baraa Mafrachi - baraawail101@gmail.com; Abdallah Al-Ani - abdallahalany@gmail.com; Amer Khalil Al Qaisi - Dr.amer_alqaisi@yahoo.com; Nadwa Bustami - Nadwa_albustami@yahoo.com

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,[1] in which ancient changes are exceptionally uncommon. Clinically, it presents with facial pain, blurred vision, headache, and seizures.[2,3,13] 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 Babre’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].

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, sensitivity to temperature, demanding anesthetic protocols, and its inability to affect extend of resection.

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 immunohistochemical 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].
DISCUSSION

Schwannomas are slow-growing tumors of the perineural Schwan cells. They may involve any myelinated nerve including peripheral, cranial, or autonomic nerves. About 25–40% of all schwannomas occur in the head and neck. 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.

Ancient schwannomas are a benign uncommon histological variant of schwannomas, first described by Ackerman and Taylor. 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 hyalination 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. 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. Ancient schwannomas involving the intracranial part of the trigeminal nerve are extremely rare and were reported in the literature only twice.

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.

Complete tumor resection with nerve function preservation, minimal postoperative morbidity and mortality are the present goals of trigeminal schwanna 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.

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 schwanna 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.
Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Ackerman LV, Taylor FH. Neurrogenous tumors within the thorax a clinicopathological evaluation of forty-eight cases. Cancer 1951;4:669-91.
2. Agarwal A. Intracranial trigeminal schwannoma. Neuroradiol J 2015;28:36-41.
3. Agrawal A, Bhake A, Meshram N, Nisha C. Ancient schwannoma of the trigeminal nerve mimicking high grade lesion. Iran J Pathol 2010;5:97-9.
4. Borges A, Casselman J. Imaging the trigeminal nerve. Eur J Radiol 2010;74:323-40.
5. Charalampidis A, Jiang F, Wilson JR, Badhiwala JH, Brodke DS, Fehlings MG. The use of intraoperative neurophysiological monitoring in spine surgery. Glob Spine J 2020;10:104S-14.
6. Gundamaneni SK, Singh M, Madhugiri VS, Rathakrishnan RK, Sasidharan GM. Trigeminal schwannoma of the sphenoid sinus--report of a rare entity. Br J Neurosurg 2014;28:281-3.
7. Kumaria A, Ingale HA, Robertson IJ, Ashpole RD. Trigeminal schwannoma presenting as a gelastic seizure: No laughing matter. Br J Neurosurg 2018:1-2.
8. Micovic MV, Zivkovic BM, Zivanovic JD, Bascarevic VL, Bogosavljevic V, Rasulic LG. Ancient olfactory schwannoma-case report and literature review. Turk Neurosurg 2017;27:656-61.
9. Neves MW, De Aguiar PH, Belsuzarri TA, De Araujo AM, Paganelli SL, Maldaun MV. Microsurgical management of trigeminal schwannoma: Cohort analysis and systematic review. J Neurol Surg B Skull Base 2019;80:264-9.
10. Puglisi G, Sciortino T, Rossi M, Leonetti A, Fornia L, Nibali MC, et al. Preserving executive functions in nondominant frontal lobe glioma surgery: An intraoperative tool. J Neurosurg 2019;131:474-80.
11. Ugokwe K, Nathoo N, Prayson R, Barnett GH. Trigeminal nerve schwannoma with ancient change. Case report and review of the literature. J Neurosurg 2005;102:1163-5.
12. Vega-Zelaya L, Pastor J. Neurophysiological monitoring techniques for the resection of malignant brain tumors located in eloquent cortical areas. Austin J Neurosurg 2015;2:1038.
13. Zhang L, Yang Y, Xu S, Wang J, Liu Y, Zhu S. Trigeminal schwannomas: A report of 42 cases and review of the relevant surgical approaches. Clin Neurol Neurosurg 2009;111:261-9.

How to cite this article: Al-Shudifat A, Mafrachi B, Al-Ani A, Al Qaisi AK, Bustami N. Ancient Schwannoma affecting the intracranial portion of the trigeminal nerve: A case report. Surg Neurol Int 2020;11:426.