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Hearing Loss Presentation and Ophthalmic Complication: Giant Trigeminal Schwannoma

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Abstract

Trigeminal Schwannoma is a very rare trigeminal nerve pathology. Variable presentations of the disease were reported in the literature. Facial pain and Trigeminal neuralgia are considered the most common presentations. Here, we are reporting a case of extra-large trigeminal lesion presented with hearing impairment which is a very uncommon presentation in small and medium sized schwannomas. Imaging studies, especially MRI plays a major role in both diagnosing and the planning of surgery. The Case was diagnosed as trigeminal schwannoma. Complete resection of the tumor was successfully achieved. Histologic testing confirmed the diagnosis. patient had a favorable postoperative course.

Keywords

Trigeminal, Schwannoma, Hearing

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1. Introduction

In 1908, Virchow has discovered a benign slow growing solitary tumor that was named as Schwannoma (neurilemmoma) as it originates from the neuroectodermal cells known as Schwan cells [1, 2]. Schwan cells are the cells that form the sheath surrounding the nerve fibers [2]. Schwannomas represents around 0.1 to 0.5 % of all brain tumors [3]. Vestibular schwannoma is the most common among all types of schwannomas [2]. Trigeminal nerve schwannoma is a rare tumor that can arise anywhere between the nerve root and the extracranial branch [2]. It accounts for 0.2 to 0.4 % of all intracranial tumors and 0.8-8% of all intracranial schwannomas [4].

Hereby, we are presenting a case which was presented with hearing impairment, investigated and found to have a large trigeminal schwannoma with intracranial and extracranial component at the middle fossa.

2. Case Presentation

A 33 years old lady not known to have any medical illnesses presented to an otolarhinology clinic in February 2018 with history of progressive hearing loss associated with tinnitus of her left ear. There was no discharge, itching or fever. She denied any history of trauma. There were no facial symptoms. Otoscopic examination pointed towards left conductive hearing loss; dull tympanic membrane on the left side and normal tympanic membrane on the right side. It was confirmed with Tympanometry and Pure tone audiometry (Figure 1). She was diagnosed with moderate conductive hearing loss in her left ear which was managed with a pressure equalizing tube (Gourmet) insertion. Six Months later, in a subsequent follow up she had the same complaint with very mild improvement. Incidentally while scoping her

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nasopharynx with a Nasopharyngoscope, a mucosal bulge was seen, therefore Computed Tomography (CT) of the brain and nasopharynx with contrast was done to rule out any nasopharyngeal pathology. Surprisingly, a huge heterogeneous 8 cm craniocaudal x 5 cm transverse x 7 cm posterior-anterior extra axial mass was seen with intracranial and extracranial component at the left middle cranial fossa with mass effect and infratemporal extension (Figure 2). She was referred urgently to Neurosurgery Department for further management.

On examination, all her motor and sensory neurological functions were intact. A Magnetic Resonance Imaging (MRI) of the brain and base of skull with Gadolinium was done and showed a large enhancing cystic hemorrhagic mass along the course of the left trigeminal nerve likely to represent Trigeminal nerve schwannoma. Moreover, a Computed Tomography Angiography (CTA) was done and showed a mass effect on the left intracranial Internal carotid artery and proximal Middle cerebellar artery.

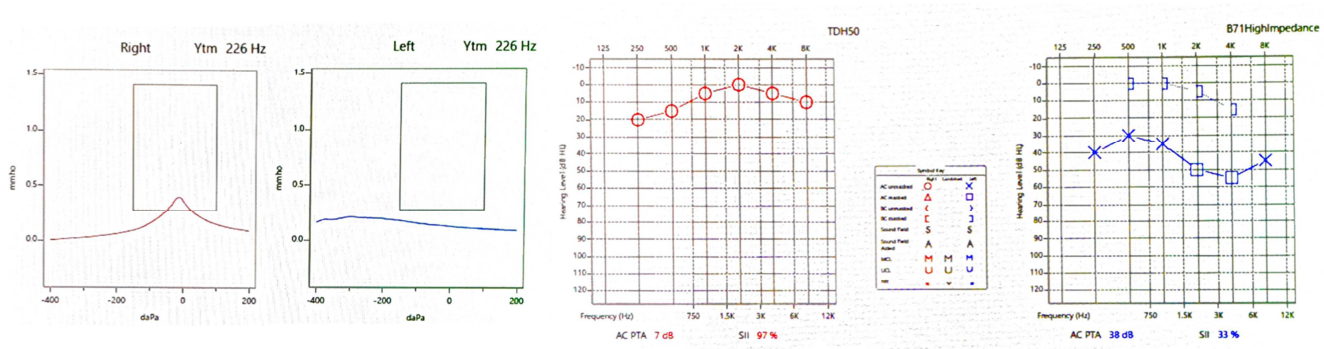

Figure 1. (A) Tympanometry: right ear represents type A and left ear represent type B. (B) Pure Tone Audiometry: Left ear conductive hearing loss. Right ear has normal sound conduction.

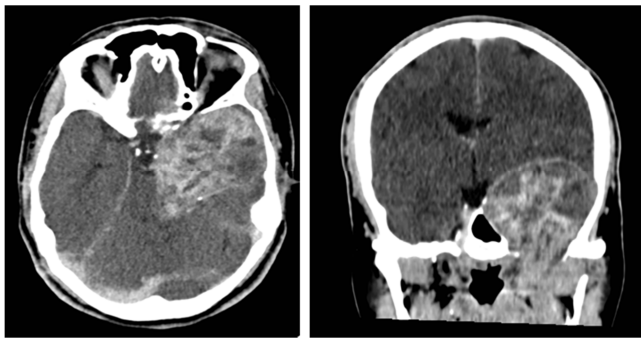

Figure 2. A large heterogeneous extra axial mass (8 cm craniocaudal x 5 cm transverse x 7 cm posterior-anterior) in the left temporal and sphenoid bones destruction and extension to the left parapharyngeal space and left cerebellopontine angle with secondary mass effect.

As the tumor was large in size, patient underwent left frontotemporal craniotomy and excision of the tumor. The tumor was located interdurally extending into the posterior fossa and the infratemporal region. The Meckel's cave and Foramen Ovale were also involved.

Postoperatively, Patient was hemodynamically stable, and her GCS was 15/15. Repeated imaging studies (Figure 4) showed no residual tumor, but there was a small right cerebellar hematoma with marked surrounding edema and adjacent small amount of subarachnoid hemorrhage. Her neurological functions were intact, except for new onset left eye esotropia. She was referred to ophthalmology clinic after 8 days of hospital stay.

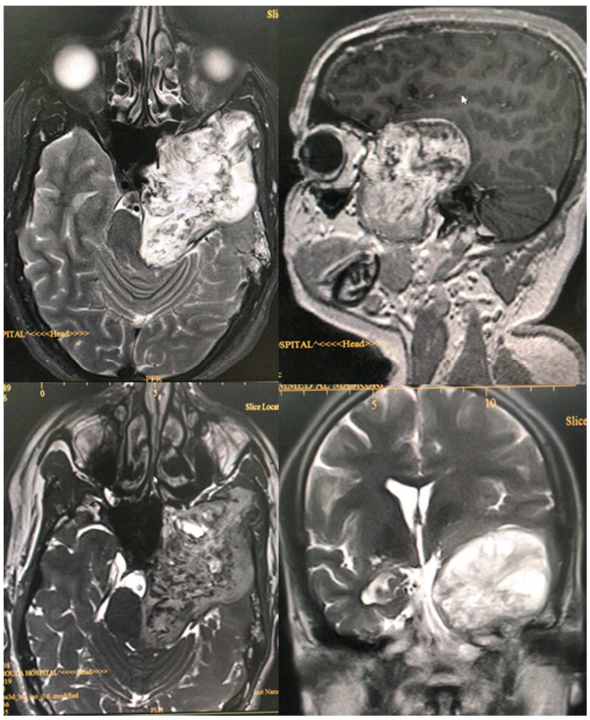

Figure 3. MRI images of the brain and base of skull with different modalities showing a large enhancing cystic hemorrhagic mass along the course of the left trigeminal nerve. The lesion is causing significant mass effect on the adjacent structures including cavernous sinus structures (left cavernous ICA), brain parenchyma with compression of the left lateral ventricle. Inferiorly, the lesion extends into the left masticator space likely through the left foramen ovale. There is a mass effect on the left side of the nasopharynx causing obstruction of the Eustachian tube opening and left mastoid effusion. There is also compression of the brainstem.

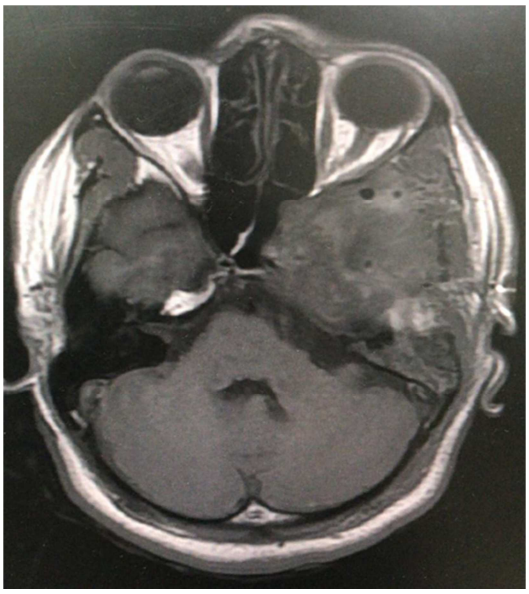

Figure 4. Axial T1 MRI image shows post-operative left frontotemporal craniotomy status. There is complete resection of the lesion. Post-operative changes include edema, air loculi and small hemorrhage are seen.

Histopathology showed typical appearance of benign schwannoma with short spindled cells with no significant cellular atypia, mitosis or coagulative tumor necrosis. The diagnosis of trigeminal schwannoma was made.

Our patient developed subarachnoid hemorrhage and edema, therefore compression against the clivus or direct damage to the nerve during tumor removal can lead to sixth nerve palsy, which was confirmed by Ophthalmology examination that favored left sixth nerve palsy. She was having diplopia on primary gaze and extraocular movement restriction towards the lateral side of the left eye (Cover test: left eye Esotropia 15°). Forced duction tests was negative. There were no signs of other cranial nerves involvement.

3. Discussion

Patients with trigeminal schwannoma can be asymptomatic for long time, however its clinical presentation varies depending on location, size and degree of tumor's extension [4]. In some cases, it presents with headache, dizziness, ataxia, hypoesthesia of the VII cranial nerve and rarely with hearing impairment [3]. Pamir and his colleagues created an algorithm for surgical management of trigeminal schwannomas [5]. According to this system, tumor size and symptoms are two major factors in the management of primary trigeminal schwannomas. Regardless of size, symptomatic tumors require surgery. Small asymptomatic trigeminal schwannomas may be treated with stereotactic radiosurgery. For residual and recurrent tumors, size is an important factor when considering management options; small masses (those less than 3 cm diameter) can be effectively treated with radiosurgery. In their articles, they reported 18 cases of trigeminal schwannomas and only two cases were tumor size was more than 6cm [5]. *Fukay etal*, reviewed 57 cases of trigeminal schwannomas and classified trigeminal Schwannoma into four categorizes according to the size; small (-1, 9cm), medium (2.0-2.9cm), large (3.0-3.9 cm) and extra large (4.0 cm) [6]. If the growth occurs in the cavernous sinus, it may lead to dysfunction of cranial nerves III, IV and VI, while growth in the prepontine cistern could lead to compressive effects on cranial nerves VII, VIII and IX [7]. In *Fukay* review, only 0.05% had hearing impairment [6]. This presentation occurs when the tumor size is extra-large and causing compression of the eight-cranial nerve. We believe that obstruction of the Eustachian tube opening is also a contributing factor for her hearing loss. There is also compression of the brainstem.

Our patient had a tumor of 8 cm in size and she presented with progressive hearing loss and tinnitus was the only symptoms that patient having for more than 6 months before the tumor found. All her motor and sensory neurological functions were intact. Hearing impairment is extremely rare presentation of trigeminal schwannoma.

Cranial nerves palsy is common complication postoperatively. *Fukay* reported sixth nerve palsy in his review 8.3% as transit and 6.3% as permanent palsy. The abducent nerve arise from horizontal sulcus between the pons and medulla and pass close to corticospinal bundles and anterior inferior cerebellar artery then through Dorello's canal, cavernous sinus and entering the orbit through superior orbital fissure [8-10]. It is long intracranial course compare to other cranial nerve course which make it vulnerable to be compressed or injured by edema, hematoma or iatrogenic during surgeries. [8-10]

4. Conclusion

Trigeminal Schwannoma is a rare tumor comprises a very small percentage of all intracranial tumors [11]. Pain, and numbness in the distribution of the fifth cranial nerve is the most common presenting complains; however, the majority is painless. Their symptoms are usually inconspicuous until the mass becomes large enough to compress the adjacent structures including lower cranial nerves [12]. The diagnosis can be confirmed by histology, however IMR findings alone can help in making the diagnose of schwannoma. Although the surgical option might be the curative treatment, it is highly individualized based on the size, location and the complexity of tumors [13, 14]. The significant evolution in microsurgeries have shown promising results in the treatment of neuroma and are expected to be the first option in future [15].

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of Interest

There are no conflicts of interest.

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