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# Voluminous Cystic Trigeminal Schwannoma: Case Report and Literature Review

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## Abstract

Voluminous trigeminal schwannomas are rare, predominantly benign growth encapsulated tumors composed of Schwann cells with controversial surgical treatment. They account for 20% to 40% of all trigeminal schwannomas and share the same imaging findings of neurinomas elsewhere. Surgery remains the treatment of choice for most lesions as long as complete excision is feasible. A 35-year-old house wife complained of a 3-month history of progressive right visual field impairment associated with headaches. Neurological examination revealed a complete cavernous sinus syndrome. CT scanning showed a voluminous cystic mass of the right parasellar compartment extending to the optic nerve and the orbit. A concomitant MRI scan revealed more exquisite anatomical details of the lesion. The patient benefitted from a subsequent CT-angiography (not included) which excluded any vascular abnormalities associated with the tumor. Surgical excision of the tumor was warranted and the patient underwent surgery a few days after admission. A pterional extradural-intradural approach combined with incision of the lateral wall of the parasellar compartment via a standard frontotemporal craniotomy was performed and pathological examination was in favor of a cystic trigeminal schwannoma. Post-operative MRI showed a complete excision of the tumor. A 2-year follow-up yielded persistent right monocular blindness. Given this typical appearance of this lesion indicative of both cisternal and parasellar compartment extension, as well as the level of controversy surrounding surgical treatment of these particular tumors; we sought to report this rare case of a voluminous cystic trigeminal schwannoma and share our humble surgical experience of dealing with these lesions. The patient was informed that non-identifying information from the case would be submitted for publication, and she provided consent.

**Keywords**

Schwannoma, Cystic, Cavernous Sinus Syndrome, Trigeminal Nerve

## 1. Introduction

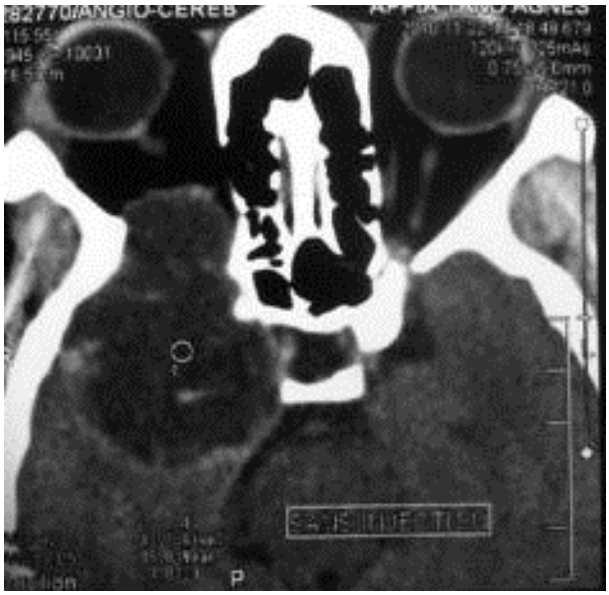
Trigeminal schwannomas are uncommon slow-growing encapsulated tumors composed of Schwann cells, which were first described by Dixon in 1846 [1]. A voluminous trigeminal schwannoma typically extends to both the cisternal and the postganglionic sections of the nerve and accounts for 20% to 40% of all trigeminal schwannomas [2]-[15]. Typically, patients would be in their third or fourth decades; and depending on the origin, the extension direction of the tumors, and the number of fibers or cells of the ganglion involved, most patients may present with clinical signs of nerve dysfunction or mass effect symptoms [5]. But in about 10% to 20% of patients, the evidence of a distinct clinical syndrome associated with voluminous trigeminal schwannoma cannot be found [5] [10] [12] [15] [16]. Trigeminal neurinomas can however, if small, arise from any portion of the trigeminal nerve namely cisternal, gasserian or the postganglionic section made of any of the three peripheral divisions of the nerve (ophthalmic, maxillary and mandibular). These tumors account for 0.07% to 0.36% of all intracranial tumors, around a third of all tumors of Meckel's cave and 0.8% to 8% of all intracranial schwannomas [6] [10] [13] [14] [15] [17] [18] [19]. As with other schwannomas, there could be an association with Neurofibromatosis type 2. Female patients are believed to be more affected than their male counterparts with a sex ratio of 1.5:1. Given the level of controversy surrounding surgical treatment of voluminous trigeminal schwannomas, we present this patient with a cystic trigeminal schwannoma treated successfully with surgical excision and share our surgical experience.

## 2. Case Report

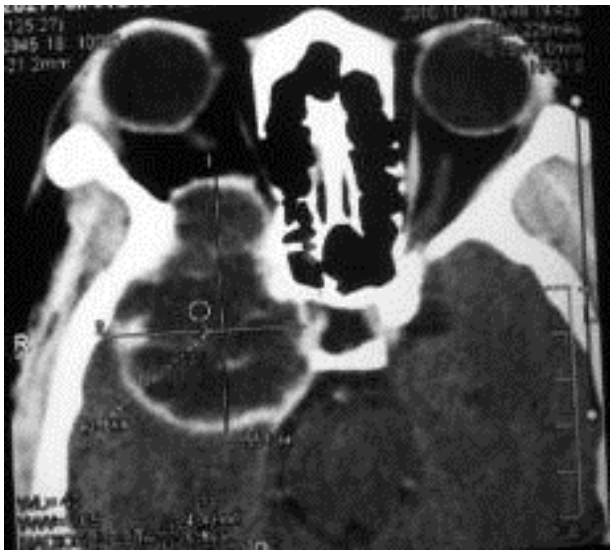
Mrs A.T, a 35-year-old right-handed house wife, with no particular medical history, complained of a 3-month history of progressive right visual field impairment associated with headaches. A thorough neurological examination revealed a fully alert patient with a complete right cavernous sinus syndrome including an axil exophthalmos, monocular blindness, hypoesthesia in the three divisions of the fifth nerve, masseter muscle wasting and palsy, third, fourth and sixth nerve palsies. The rest of the clinical examination was insignificant and the patient was otherwise well without fever.

Brain NECT scan showed a voluminous hypodense mass partially partitioned of the right parasellar compartment extending to the right optic canaland the orbit (**Figure 1(a)**). Moderate rim enhancement rather heterogeneous was observed after contrast injection (**Figure 1(b)**).

Brain MRI revealed more exquisite anatomical details of the lesion on



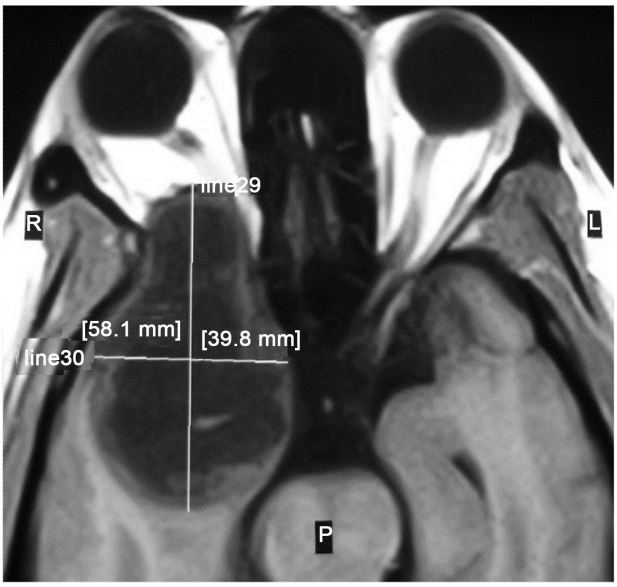
(a)



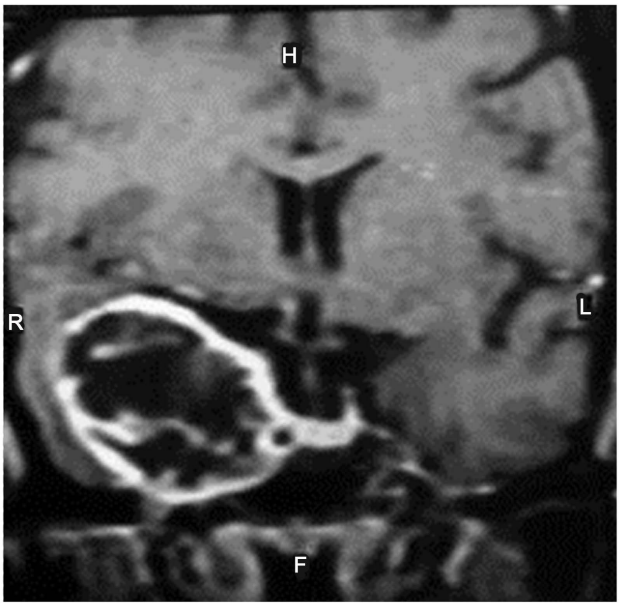
(b)

**Figure 1.** Brain NECT scan showing a voluminous dumbbell-shaped hypodense mass (58.1 × 39.8 mm) partially partitioned of the right parasellar compartment extending to the right optic canal and the orbit (a); Moderate rim enhancement rather heterogeneous was observed after contrast injection (b).

T1-weighted imaging with and without gadolinium enhancement (**Figure 2**). It is a 58.1 mm × 39.8 mm hypointense extra-axial, intradural tumor with heterogeneous enhancement and bone erosion, located in the right temporal fossa, extending into the right parasellar compartment, the right orbit and compressing the right optic nerve. It is undoubtedly supratentorial, free from the carotid artery and Meckel's cavum. Differential diagnosis for these lesions included a voluminous cystic trigeminal schwannoma, atypical epidermoid cyst of the temporal fossa, meningioma, cystic lymphangioma or cholesteatoma. A subsequent




(a)




(b)

**Figure 2.** T1W MRI imaging showing a 58.1 mm × 39.8 mm hypointense extra-axial, intradural tumour with heterogeneous enhancement and bone erosion, located in the right temporal fossa, extending into the right parasellar compartment, the right orbit and compressing the right optic nerve. The tumour is supratentorial, free from the carotid artery and Meckel's cave.

CT-angiography (not included) excluded any vascular abnormalities associated with the tumor.

Surgical excision of the tumor was warranted and the patient underwent surgery a few days after admission. A pterional extradural-intradural approach combined with incision of the lateral wall of the parasellar compartment via a standard frontotemporal craniotomy was performed.

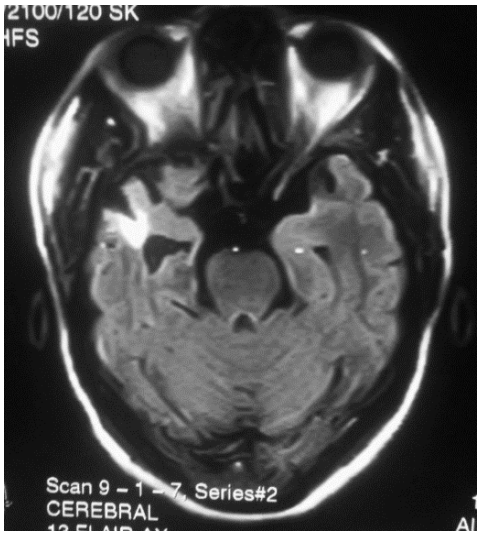
The patient was positioned supine with slight neck extension, the head turned 30˚ to the left and secured with a Mayfield three-point skull fixation. A generous question mark skin incision was fashioned starting at the level of the zygoma within 1 cm anterior to the tragus toward the midline and ending just behind the hairline. After retracting the skin flap anteriorly, careful interfascial dissection of the temporalis muscle was carried out to protect the temporal branch of the facial nerve. The superficial temporalis fascia was incised 3 cm posterior to the frontal process of the zygomatic bone and reflected with the skin flap. Further dissection was carried out to expose the frontal, temporal and zygoma. A standard frontotemporal craniotomy was performed. After removing the bone flap, and beginning at the frontal limit of the craniotomy, an S-shaped dural incision was carried to the level of the impression of the lesser sphenoid wing. After opening the dura and further dissecting, drainage of the chiasmatic cistern allowed maximal brain relaxing. The superior orbital fissure was exposed after removing about 1 cm of the lateral orbital roof with a high-speed drill and rongeurs. Dissection was then extended into the middle cranial fossa. The anterior clinoid process was removed extradurally and we further achieved intra-extradural exposure of the lateral parasellar compartment. The tumor appeared cystic filled with a clear brownish fluid and was free from adjacent vascular structures. Under microscope, drainage of the cyst was achieved and the rest of the tumor was excised progressively in pieces until deemed macroscopically complete.

The post-operative course was uneventful and the patient had a significant improvement of her symptoms. Post-operative MRI showed a complete excision of the tumor (**Figure 3**). But a 2-year follow-up yielded persistent right monocular blindness.

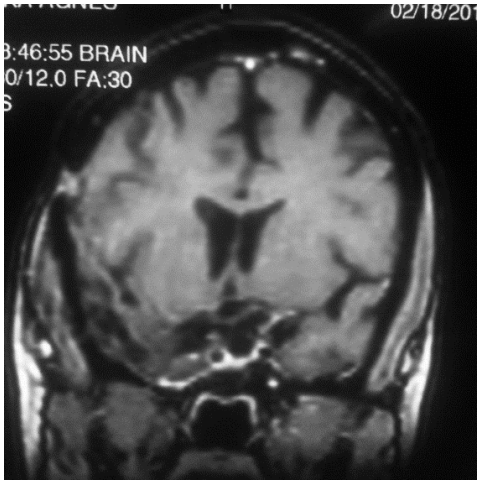
Pathological examination of the specimen revealed spindle-shaped Schwann cells often swirling or entangled, the cytoplasm of which was eosinophilic, containing a rounded or elongated hyperchromatic nucleus with moderate anisocaryosis and no mitotic activity. Along with cysts, hemorrhage and edema, scanty nuclear palisading without Verocaybody formation was also seen. The stroma contained fibrous and hyalinised blood vessel walls. This histological aspect is consistent with Antoni B cystic trigeminal schwannoma (**Figure 4**).

## 3. Discussion

Trigeminal schwannomas are uncommon slow-growing encapsulated tumors composed of Schwann cells. They account for 0.07% to 0.36% of all intracranial tumors and 0.8% to 8% of all intracranial schwannomas [10] [14] [17] [19]-[25]. Interestingly, about 30% of trigeminal neurinomas are totally or predominantly confined to any portion of the trigeminal nerve namely cisternal, gasserian or the postganglionic section made of any of the three peripheral divisions of the nerve (ophthalmic, maxillary and mandibular) [2]-[15]. They can however, in less than 20% be voluminous with significant tumor extension in both the middle and posterior fossa [1] [5] [21]. Given that most authors with substantial se-




(a)




(b)

**Figure 3.** Postoperative MRI scan showing complete tumour removal.

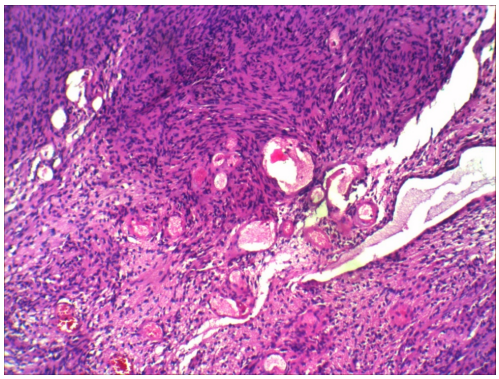


**Figure 4.** (HE × 100): Histological aspect of Antoni B tumour cells showing entangled spindle-shaped Schwann cells with eosinophilic cytoplasm and elongated hyperchromatic nucleus with moderate anisocaryosis and no mitotic activity. Along with cysts, haemorrhage and oedema, scanty nuclear palisading without Verocay body formation can be seen. The stroma contains fibrous and hyalinised blood vessel walls.

ries report more caudal extensions [1] [5] [7] [11] [21]; our patient had a rather unusual anterior extension to the orbit with significant compression of the optic nerve which makes this case relevant. It is the authors' view that patients and healthcare professionals alike should seek neurosurgical referral as soon as it is feasibly possible, because late neurosurgical care in these particular instances would increase the likelihood of irreversible visual impairment.

It is believed that comprehension of the biologic behavior, clinical features and to a certain extend appropriate surgical approach for removal of these tumors is facilitated by an understanding of the anatomical course of the trigeminal nerve from the posterior fossa to the middle fossa. The trigeminal nerve is intradural in the posterior fossa and becomes interdural when it enters the Meckel cave. As it prepares to fan out into three divisions, the gasserian ganglion lays extracarvernously alongside the mandibular branch, whereas the ophthalmic and maxillary branches are intracavernous [2] [9] [26]. Voluminous trigeminal schwannomas are believed to arise as argued by Goel *et al.* at the site of entry of the nerve into the Meckel dural cave, where the rigid bone and the dural matter at the petrous apex confine tumor growth, allowing tumor spreading from both direction, creating this narrow connection between both tumor extremities typical feature of these schwannomas, by which voluminous trigeminal neurinomas can be differentiated from other neoplasms in this area [7].

Typically, patients would be in their third or fourth decades and; depending on the origin, the extension direction of the tumors, and the number of fibers or cells of the ganglion involved, most patients may present with clinical signs of nerve dysfunction or mass effect symptoms [5] which happen to be the case of our patient. But in about 10% to 20% of patients, the evidence of a distinct clinical syndrome associated with voluminous trigeminal schwannoma cannot be found [5] [10] [12] [15] [16]. Peculiarly, a size-effect relationship between the dysfunction of the nerve and the size of the tumor has not been found. As with other schwannomas, there could be an association with Neurofibromatosis type 2 [27]. Female patients are believed to be more affected than their male counterparts with a sex ratio of 1.5:1 [27].

Given that evidence of distinct clinical syndromes associated with voluminous trigeminal schwannoma cannot be found, neuroimaging investigations, therefore, are major diagnostic modalities. Computed tomography and MRI are supplementary for each other and should be consider gold standard investigative modalities for the diagnosis of voluminous trigeminal neurinomas [1] [5]. The typical appearance on CT scan is that of a voluminous masse, which is slightly hypodense and enhances heterogeneously with contrast if cystic. MRI scanning demonstrates often more exquisite anatomical details of the lesion with low signal intensity on T1W and high signal intensity on T2W images and; heterogeneous enhancement after gadolinium infusion if cystic, as was the case for our patient.

Yoshida and Kawase divided trigeminal schwannomas into 3 categories according to their locations and, Goel added 2 more categories recently [7] [21].

They are middle fossa type, posterior fossa type, dumbbell-shaped type, extracranial peripheral type, and mixed type trigeminal neurinomas. Ramina *et al.* proposed a more extensive classification which assures the best surgical approach for each type of the lesion and classifies the level of difficulty according to the surgical challenge [28]. Our patient had a type B trigeminal schwannoma which stands in the middle in the order of surgical difficulty.

Angiography is now seldom used except in cases where vascular abnormalities or an intracranial aneurysm should be ruled out.

Definite diagnosis of cystic trigeminal schwannoma is made after pathology analysis. Two types of schwannoma cell tumors are often described in most pathology reports. Antoni type A are fibrillary, intensely polar and elongated appearing tissue type, and have closely packed elongated nuclei which demonstrate a thickened middle portion with tapered ends and resemble the wooden spindles used in textile spinning and are associated with Verocay bodies. Antoni B type tumor cells have, as was the case of our patient, spindle-shaped Schwann cells often swirling or entangled, the cytoplasm of which is eosinophilic, containing a rounded or elongated hyperchromatic nucleus with moderate anisocaryosis and no mitotic activity. Along with cysts, hemorrhage and edema, scanty nuclear palisading without Verocaybody formation is also seen. The stroma contains fibrous and hyalinised blood vessel walls.

These tumors are predominantly benign growth encapsulated schwannomas and complete surgical removal when feasible should be the gold standard treatment option. Surgical treatment of voluminous trigeminal schwannomas remains controversial. Recent advances in microsurgery and various skull base approaches have led to improved outcomes and a high rate of resection of 77% to 100% [2]-[15] [17] [18] [19] [21] [22] [23] [24] [25] [28]. Our post-operative MRI scan demonstrated complete resection of the tumor. Intradural skull base approaches include frontotemporal transtentorial, subtemporal, suboccipital or retromastoid routes performed either as part of a combined approach or as a separate operation. Each approach is chosen based on the anatomy of the tumor. We performed a pterional extradural-intradural approach combined with incision of the lateral wall of the parasellar compartment via a standard frontotemporal craniotomy. Zhou *et al.* favored an extraduro transduro transtrigeminal pore approach via frontotemporal craniotomy and orbitozygomatic osteotomy with varying outcomes [5]. Surgical morbidities or unimprovement and worsening of cranial nerves after surgery are not linear from one study to the other [1] [5]. The recurrent rates after total tumor removal reported in the literature are 2.5% to 14% [1] [9]. Generally speaking, improved surgical results and outcomes are believed to be attributed to an accumulation of surgical experience, improvement of techniques, and the use of MRI and skull-base approaches. The ultimate goal in the surgery of voluminous trigeminal schwannomas is complete removal of the tumor. However, in cases where the tumor capsule is heavily adherent to important neurovascular structures, it is advisable to abandon complete removal rather than risking damage to important neurovascular structures.

In these instances, radiosurgery using gamma knife might be required in addition to subtotal excision, to control potential long-term tumor regrowth [1] [5].

## 4. Conclusion

It is evident from this extensive literature review that small and middle-sized trigeminal neurinomas are best dealt with radiosurgery. However, voluminous trigeminal schwannomas are candidates for microsurgery with skull-base approach, with radio surgery as an adjuvant option for residual or recurrent tumors that cannot surgically be removed or for patients unable to undergo surgery.

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