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Trigeminal Schwannoma with Extracranial Extension and Brainstem Compression

Andrew J. Coniglio, MS2¹, Matthew C. Miller, M.D.², Kevin A. Walter, M.D.³, and Benjamin T. Crane, M.D., Ph.D.²

¹University of Rochester School of Medicine and Dentistry, Rochester, NY

²University of Rochester-Strong Memorial Hospital, Department of Otolaryngology, Rochester, NY

³University of Rochester-Strong Memorial Hospital, Department of Neurosurgery, Rochester, NY

Trigeminal schwannomas are rare tumors that comprise about 0.2% of all intracranial tumors (1,2,4,5). They are predominately benign growths and prognosis is usually good due to improvements in diagnostic imaging and surgical excision (1,2). Women are affected slightly more than men and most cases present in the third or fourth decades (1,2). Patients present most commonly with facial hypoesthesia, pain, and paresthesia as well as diplopia and other visual disturbances (1,2,5). Wasting of the temporalis and pterygoid muscles is also common. Less frequent symptoms include hearing loss, ataxia, and dysarthria as well as pathological crying thought to be caused by brainstem compression (1). The tumor size tends to be large because the mild symptoms make early diagnosis difficult.

Trigeminal schwannomas may originate from the trigeminal dorsal root entry zone, the Gasserian ganglion, or any of the three trigeminal nerve branches, which can lead to complex tumors that occupy multiple fossae (1). In an attempt to simplify tumor presentation, Jefferson (3) grouped trigeminal schwannomas into three types based on their location within the cranium. Type A denotes tumor growth in the middle fossa while type B constitutes tumor in the posterior fossa anterior to the brainstem. Type C includes both middle and posterior fossa and is referred to as a “dumbbell” type. Added later, Type D is used if the tumor has an extracranial extension.

Similarly, Yoshida et al. (5) classified trigeminal schwannomas in hopes of unifying a surgical approach based on presentation. M is used for a tumor found in the middle fossa, P for posterior fossa and E for an extracranial extension. Extracranial tumors were further divided into orbital, infratemporal, and pterygopalatine fossae extensions. With this system, six classifications are possible. A “dumbbell” tumor extending in both the middle and posterior fossa, a common growth formation, is classified as MP.

A 30-year-old male presented with a 6-month history of right-sided facial pain in the V-2 distribution and swelling which he initially believed to be a dental infection. He also reported at least a 10-year history of occasional right eye “twitching” and intermittent facial paresthesia for which he had not previously sought treatment.

A facial CT scan was obtained which revealed a defect in the base of skull along the middle cranial fossa with possible encephalocele displacing the posterior wall of the maxillary sinus (Figure 1). MRI revealed a large, well-defined, lobulated, heterogeneous mass lesion along

the right trigeminal nerve. The mass compressed the brainstem with displacement of the pons and midbrain. There was involvement of Meckel's cave with marked widening of his right foramen ovale. The mass extended into the parapharyngeal space and displaced the parotid outward with remodeling of the posterior wall of the right maxillary sinus (Figure 2).

A stereotactic biopsy was obtained of the middle cranial fossa portion of the tumor via the middle temporal gyrus. This biopsy revealed low-grade spindle cell neoplasm consistent with schwannoma.

Based on the radiological findings, this very rare trigeminal schwannoma involves the middle cranial fossa, posterior fossa, and parapharyngeal space. Tumors involving these 3 represent <10% of trigeminal schwannomas in large case series (1,2). While cases with brainstem compression have been noted (1), there has thus far been no published report of a tumor with extracranial extension and brainstem compression. This case demonstrates that these tumors can grow quite large before becoming symptomatic. The patient has undergone resection of the intracranial portion and is currently awaiting staged resection of parapharyngeal component.

The tumor was removed using a staged approach. The initial stage was a combination suboccipital and middle fossa approach with complete removal of the intracranial portion of the tumor. The dura defect was closed using multiple bovine paracardial grafts. Three months later, a second stage via a mandibular swing approach was performed to address the residual parapharyngeal space component of the tumor with a gross total resection. The skull base defect from the parapharyngeal approach was managed with a muscle graft and biomaterial based dural sealant product. After surgery the patient had continued right sided facial pain and numbness with his cranial nerve deficit limited to the trigeminal nerve.

References

1. Fukaya R, Yoshida K. Trigeminal Schwannomas: Experience with 57 Cases and a Review of the Literature. *Neurosurgical Review*. 2010; 34.2:159–71. [PubMed: 20963463]
2. Goel A, Muzumdar D, Raman C. Trigeminal neuroma: analysis of surgical experience with 73 cases. *Neurosurgery*. 2003; 52:783–790. [PubMed: 12657173]
3. Jefferson G. The trigeminal schwannomas with some remarks on malignant invasion of the gasserian ganglion. *Clin Neurosurg*. 1955; 1:11–54. [PubMed: 14379464]
4. Pollack IF, Sekhar LN, Jannetta PJ, Janecka IP. Neurilemmomas of the trigeminal nerve. *J Neurosurg*. 1989; 70:737–745. [PubMed: 2709114]
5. Yoshida K, Kawase T. Trigeminal schwannomas extending into multiple fossae: Surgical methods and review of literature. *J Neurosurg*. 1999; 91:202–211. [PubMed: 10433308]

Figure 1.

CT imaging using 0.625-mm contiguous sections in the axial (A) and coronal (B,C) revealed a significant defect with a soft tissue mass protruding from the middle cranial fossa through a widened foramen ovale (*arrow*) extending into the parapharyngeal space.

Figure 2.

MRI revealed a lobulated, heterogenous mass lesion extending 9.2 cm along the course of CN V, and 8.0 cm in the dorsal-ventral direction. *A*, Gadolinium enhanced T1-weighted coronal image showing the intracranial extension of the tumor through the foramen ovale. *B*, T2- weighted axial image showing marked compression of the brainstem (*arrow*). *C*, T2-weighted axial image demonstrating anterior extent of the tumor. *D*, Gadolinium enhanced T1-weighted sagittal image demonstrating extension through the foramen ovale.