case report

Multicompartmental Trigeminal Schwannomas: Dumbbell Tumors Revisited

abstract

Multicompartmental trigeminal schwannomas (MTSs) are a rare and complex but treatable group of tumors. Herein, we describe the clinicoradiologic presentation of two patients with MTS. The two illustrated distinct case reports highlight the role of imaging and the outcome of two different types of MTS. The Discussion summarizes the literature to date, which will help the reader diagnose these tumors in a timely manner and manage them appropriately.

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CASE 1

A 40-year-old woman presented with progressively increasing paresthesia and numbness along the left side of the face. She also complained of on-and-off headaches and ataxia. Weakness of muscles of mastication was noted on clinical examination. Contrast-enhanced magnetic resonance imaging (MRI) of the brain was performed, which revealed a large posterior fossa mass lesion with an extra-axial component on the left side measuring $5.9 \times 4.1 \times 3.3$ cm in anteroposterior, craniocaudal, and transverse dimensions, respectively. The lesion was hypointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and showed intense postcontrast enhancement with few peritumoral cystic areas. There was widening of the left cerebellopontine angle cistern and displacement of the left half of the midbrain and pons to the right side with significant mass effect. The lesion extended anteriorly into the lateral compartment of the left cavernous sinus up to the left Meckel cave and encased the cavernous portion of left internal carotid artery (Fig 1). On the basis of the clinical presentation and the imaging characteristics, a provisional diagnosis of schwannoma was made, and the tumor was excised through a left subtemporal approach. The histopathology revealed a nerve sheath tumor with predominant Antoni A areas with few (< two) mitotic figures. On immunohistochemistry, the tumor was diffusely positive for S100 protein (cytoplasmic). The histopathology was consistent with benign schwannoma. The final diagnosis was multicompartmental trigeminal schwannoma (MTS) type E as per Ramina et al[1] and type MP as per Yoshida and Kawase[2] classification. Immediate postoperative imaging revealed no residual disease, and follow-up imaging 1 year after surgery showed no recurrent disease.

CASE 2

A 22-year-old woman presented with chronic left facial pain and numbness along divisions of the left trigeminal nerve. Multiplanar MRI of the neck and brain was performed that revealed an extra-axial mass in the left infratemporal fossa and masticator space extending intracranially up to the left Meckel cave. The lesion was seen to extend superiorly via an enlarged foramen ovale into the parasellar and left cavernous region. The lesion was relatively isointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and showed relatively homogenous postcontrast enhancement (Fig 2). The classic clinical presentation with characteristic anatomic location and features on MRI suggested the diagnosis of trigeminal (mandibular division) nerve schwannoma. The tumor was excised through a transmaxillary approach. The histopathology of the lesion revealed a nerve sheath tumor composed of Antoni A cells that were strongly positive for S100 protein on immunohistochemistry. The histopathologic diagnosis was cellular variant of schwannoma (benign). The final diagnosis was MTS type A as per Ramina et al[1] and type ME as per Yoshida and Kawase[2] classification. Immediate postoperative imaging revealed no residual disease, and follow-up imaging at 2 years showed no recurrence.

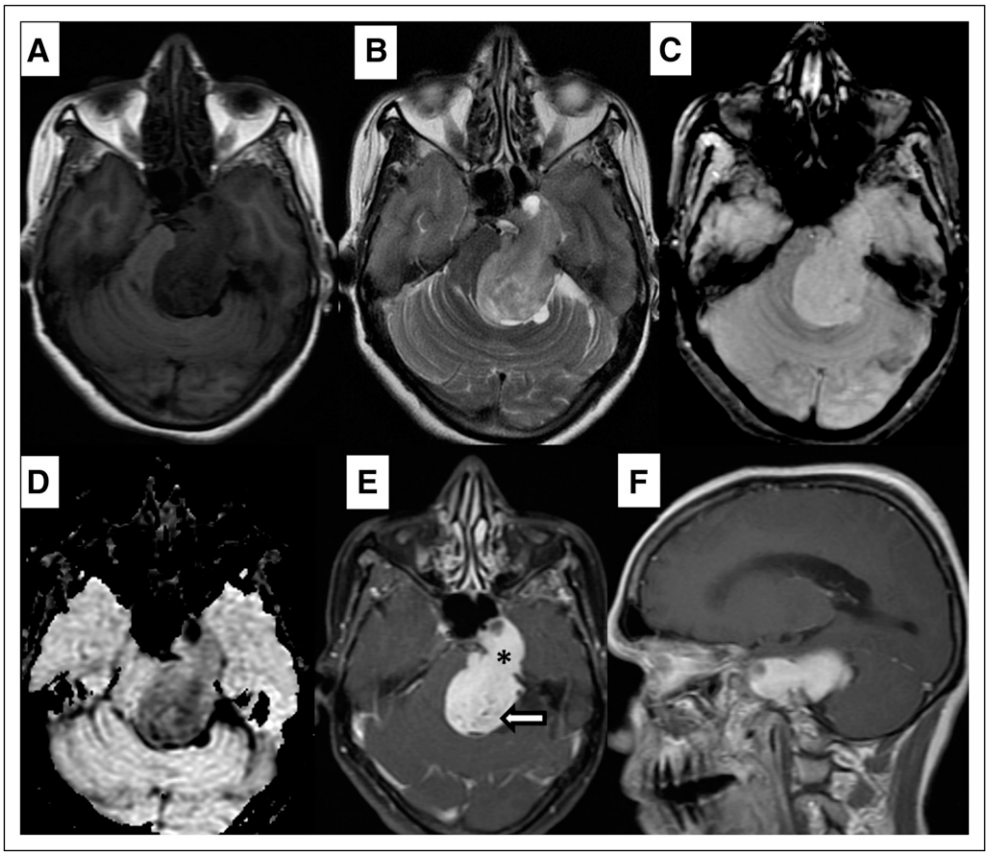

Fig 1 –
Axial (A) T1-weighted and (B) T2-weighted images showing a large posterior fossa mass lesion with extra-axial component on the left side, causing widening of the left cerebellopontine angle cistern. The mass was hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images. The mass abutted the left half of the midbrain and pons, displacing the brainstem to the right side and causing mass effect on the brainstem. The lesion extended anteriorly into the left cavernous sinus, encasing the cavernous portion of left internal carotid artery. No intralesional hemorrhages were noted on (C) gradient recalled echo images, and (D) exponential apparent diffusion coefficient imaging showed facilitated diffusion in the mass. (E and F) Postcontrast T1-weighted images showed intense postcontrast enhancement with few peritumoral cystic areas. The involvement of preganglionic segment (arrow) and postganglionic segment (*) is depicted in part E. The characteristic anatomic location, extent, and signal pattern suggested the diagnosis of multicompartmental trigeminal schwannoma type E as per Ramina et al[1] and type MP as per Yoshida and Kawase[2] classification.

DISCUSSION

Benign schwannomas of the trigeminal nerve are less common than acoustic neuromas, accounting for 0.8% to 8% of all schwannomas and 0.2% to 0.4% of all intracranial tumors. They primarily arise in the gasserian ganglion, with the cell of origin being the Schwann cell.[3-6] These tumors are slow growing and most often clinically present with facial pain that is typically described by the patient as burning in nature. When the tumor enlarges to involve the motor division of the nerve, motor dysfunction of the muscles of mastication may occur. Involvement of the sensory division of the nerve may results in sensory paresthesias and diminished corneal reflex. Growth within the cavernous sinus leads to dysfunction of oculomotor, trochlear, and abducent nerves, and enlargement within the cerebellopontine angle cistern can cause clinical symptoms related to compressive effects on facial, vestibulocochlear, and glossopharyngeal nerves.[3,6]

The normal trigeminal nerve exits the pons anterolaterally and courses through the prepontine cistern to enter the Meckel cave, a dural invagination along the medial aspect of the middle cranial fossa. The ophthalmic and maxillary divisions of the trigeminal nerve course anteriorly through the lateral wall of the cavernous sinus, whereas the mandibular division proceeds inferiorly via the foramen ovale.[4,5] The fifth nerve is primarily sensory to the face except for the motor root that does not enter the ganglion and joins the mandibular division beneath the foramen ovale.[4,5] Three major types of trigeminal schwannomas have been described based on the origin, as follows: in the preganglionic segment presenting as a dumbbell-shaped posterior fossa mass; within the gasserian ganglion presenting as a dumbbell-shaped Meckel cave mass; and in the postganglionic branches presenting as an infratemporal mass.[6,7] Depending of the origin, these tumors are either unicompartmental or multicompartmental. Various classifications that have been used for categorizing these tumors include the following: the Jefferson[8] classification, which is based on the anatomic origin of the tumor; the classification by Samii et al[7]; and the Yoshida and Kawase[2] classification, which is based on the tumor extent (Table 1). Ramina et al[1] modified the classification by Samii et al[7] and categorized these tumors into six types based on radiologic findings that have implications in planning the surgical approach for these tumors (Table 2).

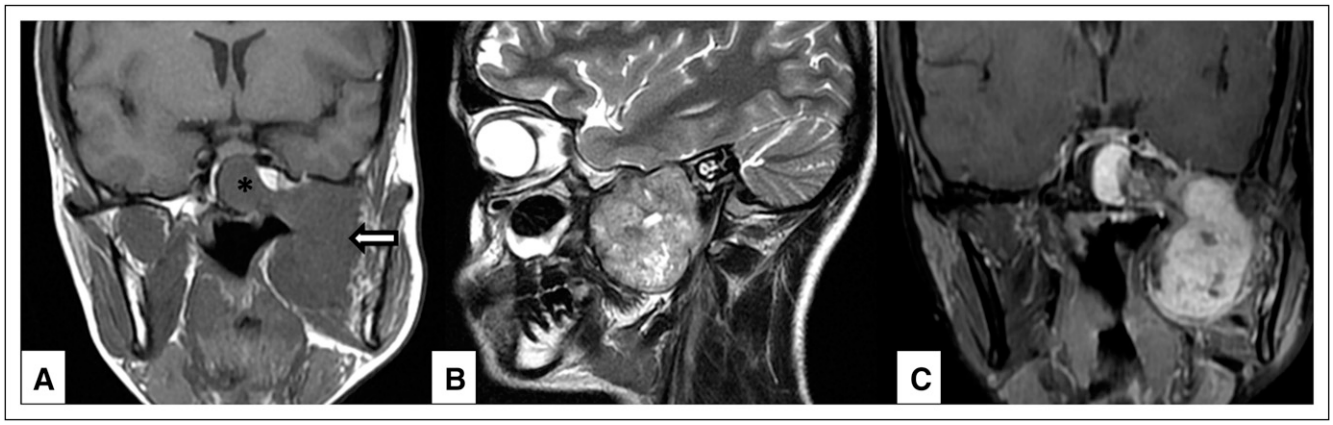

Fig 2 –
(A) Coronal T1-weighted image showing an isointense to hypointense mass in the left temporal fossa in the region of left Meckel cave (*) that extends inferiorly via an enlarged foramen ovale into the infratemporal fossa and left masticator space (arrow). The lesion was (B) heterogeneously hyperintense on sagittal T2-weighted images and (C) showed relatively homogenous enhancement on postcontrast T1-weighted images. The final diagnosis was multicompartmental trigeminal (mandibular division) schwannoma type A as per Ramina et al[1] and type ME as per Yoshida and Kawase[2] classification.

Preoperative imaging with computed tomography (CT) or MRI has significantly improved the surgical outcome of these tumors.[6] On imaging, these lesions are typically multicompartmental dumbbell-shaped masses composed of a cisternal and a cavernous sinus component. However, small unicompartmental masses confined to one section of the nerve may be seen.[4,5,9] The radiographic findings of a trigeminal schwannoma depend on the location of the tumor and may include erosion of the dorsum sellae or petrous apex and enlargement of outlet foramina such as the foramen ovale, foramen rotundum, or superior orbital fissure. Skull radiographs may reveal these osseous changes if the tumor is large.[3] On unenhanced CT imaging, these lesions are usually isodense but can also reveal variable attenuation. These lesions tend to show homogenous enhancement on postiodinated contrast imaging. In multicompartmental tumors, bone algorithm CT imaging further helps in assessment of the bony margins and status of the foramen.[4,5,10]

Contrast-enhanced MRI is preferred over CT imaging because of multiplanar capability and absence of Hounsfield artifact from the skull base. MRI provides exquisite anatomic details and characteristic tissue signal intensity patterns that are helpful in differentiating the primary tumors of the trigeminal nerve, Meckel cave, and cavernous sinus. MRI also helps in the evaluation of the extent of tumor for preoperative planning.[4,10,11] In addition, MRI is sensitive for detection of additional neuromas, which is a consideration in patients with neurofibromatosis type 2.[4,6,10] The lesions typically appear isointense to brain parenchyma on T1-weighted images (cystic areas, if present, appear hypointense). On T2-weighted images, they show hyperintense signal intensity compared with brain parenchyma (cystic areas appear hyperintense). Because they are extra-axial in location, these tumors can have a CSF cleft along their margins, which on T2-weighted images is seen as a sharp high hyperintense signal between tumor and the adjacent parenchyma. On postcontrast images, trigeminal schwannomas show marked homogenous enhancement; however, they may show heterogeneity as a result of presence of necrotic or cystic areas.[4,5,10] Most of these schwannomas show higher signal both on diffusion-weighted images and apparent diffusion coefficient mapping, which is a result of the T2 shine-through effect and not a result of cellular diffusion restriction. However mean apparent diffusion coefficient maps in these lesions show high signal intensity, suggesting facilitated diffusion and, thus, benign tumor biology.[4,12]

The differential diagnoses of these lesions depend on the size and location of the tumor. When the lesion is large and extends into the cerebellopontine angle, the differential diagnoses include acoustic schwannoma, meningioma, ependymoma, metastasis, and chondrosarcoma. The common differential diagnoses for smaller lesions confined to the Meckel cave include pituitary macroadenomas, internal carotid artery aneurysms, and vascular

Table 1 – Tumor Extent–Based Classification of Trigeminal Schwannomas

Samii et al[7]	Yoshida and Kawase[2]	Radiologic Tumor Extent
Type A	M	Intracranial tumor predominantly in the middle fossa
Type B	P	Intracranial tumor predominantly in the posterior fossa
Type C	MP	Intracranial dumbbell-shaped tumor in the middle and posterior fossa
	E	Extracranial space
	ME	Middle fossa and extracranial space
Type D	MPE	Extracranial tumor with intracranial extensions

Table 2 – Modified Ramina et al[1] Classification of Trigeminal Schwannomas and Management Implications

Category	Radiologic Tumor Extent	Surgical Approach
Type A	Predominantly extracranial tumor with small extension in the middle fossa	Extradural transmaxillary approach or transmaxillary ± extradural middle fossa approach
Type B	Intracranial tumor predominantly in the middle fossa with extracranial extension	Intradural middle fossa approach; large extradural tumor extensions through transmaxillary approach
Type C	Tumor in the middle fossa	Middle fossa approach either extradural or intradural approach: intradurally (for large lesions) or extradurally (for small lesions)
Type D	Tumor in the posterior fossa	Retrosigmoid approach
Type E	Tumor with middle and posterior fossa extensions	Presigmoid approach or two-stage retrosigmoid and middle fossa approach
Type F	Tumor with extracranial, middle, and posterior fossa extensions	Combination of above-mentioned approaches

malformations.[4,5,12] Although presence of classic clinical presentation and imaging features suggests the diagnosis, tissue diagnosis is required for confirmation and detection of malignant-variant MTS. On microscopy, the characteristic features include Antoni A cells, polyhedral cells with large pleomorphic hyperchromatic nuclei, and plump spindle cells. On immunohistochemistry, the most important markers that help differentiate schwannomas from other spindle cell tumors include S100 protein, myelin basic protein, and leucine.[12,13]

Our patient cases represent two distinct types of dumbbell-shaped MTS—one having a predominant posterior fossa with a middle cranial fossa component and the second tumor predominantly being extracranial with a small middle cranial fossa component. Both tumors had classic clinical presentation and MRI morphology that were suggestive of trigeminal nerve involvement with working diagnosis of schwannomas. The tumors were completely resected with excellent surgical outcome. To conclude, MTSs are rare tumors that not only pose a diagnostic dilemma, but also a surgical challenge. MRI is the imaging modality of choice for characterizing these tumors and plays a crucial role in planning the surgical approach.

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AUTHORS' DISCLOSURES OF POTENTIAL CONFLICTS OF INTEREST

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