

Expansile Traumatic Neuroma of the Intratemporal Facial Nerve

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Abstract

Objectives To present a rare case of traumatic facial neuroma involving the geniculate ganglion and review relevant literature.

Patient Thirty-year-old man.

Intervention Microsurgical resection via combined mastoid-middle fossa approach with great auricular nerve interpositional graft.

Main Outcome Measures Patient demographics and pre- and postoperative facial nerve function.

Results A 30-year-old man with a reported history of prior Bell's palsy developed progressive complete (House–Brackmann VI) right facial paralysis following blunt trauma. Imaging was strongly suggestive of a geniculate ganglion hemangioma. As the patient had no spontaneous improvement in his poor facial function over the course of 9 months, he underwent resection of the facial nerve lesion with great auricular nerve graft interposition via a combined mastoid-middle fossa approach. Histopathology demonstrated disorganized fascicles, with axonal clustering reminiscent of sprouting/regeneration following trauma. No cellular proliferation or vascular malformation was present.

Conclusion Traumatic facial nerve neuromas can occur following temporal bone trauma and can closely mimic primary facial nerve tumors. Akin to the management of geniculate ganglion hemangioma and schwannoma, preoperative facial function largely dictates if and when surgery should be pursued.

Keywords

- ▶ facial nerve
- ▶ traumatic neuroma
- ▶ facial nerve schwannoma
- ▶ geniculate
- ▶ hemangioma

Introduction

Traumatic neuroma (TN) is a nonneoplastic proliferation of a peripheral nerve.¹ The size of the TN is inversely proportional to the extent of healthy neural regeneration during re-establishment of nerve continuity.² TN are composed of

disorganized nerve fascicles, Schwann cells, and endoneurial and perineurial fibroblastic cells in a dense collagenous matrix.² Facial nerve TN is rare and can develop following extensive injury or more subtle trauma that does not result in nerve discontinuity. It may also develop in association with chronic middle ear inflammation.³ These lesions may occur

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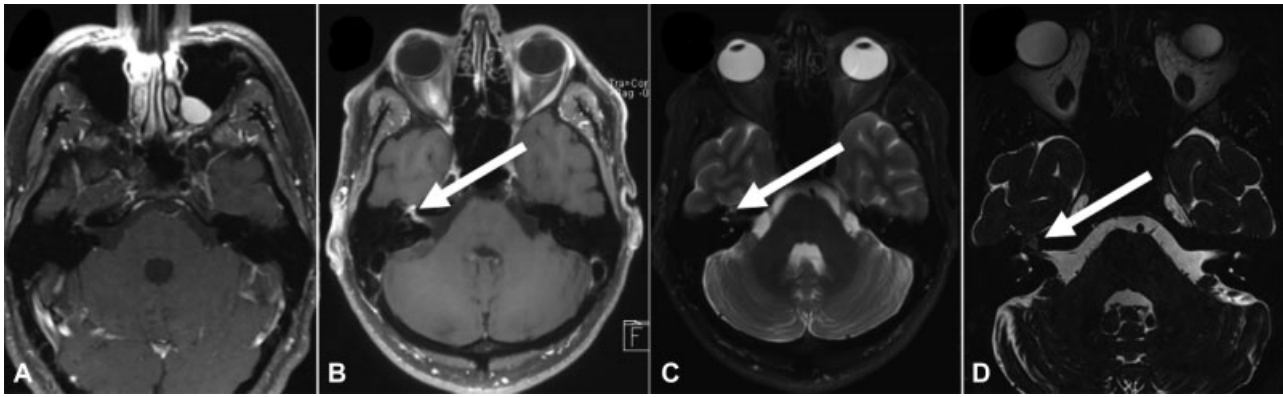


Fig. 1 (A) Postcontrast T1-weighted MRI 2.8 years before presenting to our center demonstrating no evidence of lesion. (B) Recent postcontrast T1-weighted (C) T2-weighted fast spin echo, and (D) thin slice heavily T2-weighted MRI demonstrated an expansile mass involving the right meatal, labyrinthine, geniculate, and proximal tympanic facial nerve.

in any segment of the facial nerve, but most commonly involve the labyrinthine, geniculate, and tympanic segments.^{4,5} In this report, a case of facial nerve TN following blunt trauma is presented, along with imaging, histopathology, and management presented in a surgical video.

Case Report

A 30-year-old man presented to the authors' institution with complete right-sided facial paralysis following a downhill skiing injury. More remotely, he was diagnosed with Bell's palsy 3 years earlier and was treated with corticosteroids with incomplete recovery. Imaging at that time did not demonstrate any abnormality of the facial nerve. The skiing accident occurred 9 months prior to presentation to our

institution. During evaluation, he endorsed mild imbalance and denied hearing loss. Examination demonstrated normal cranial nerve function with the exception of House-Brackmann (HB) grade VI facial function. Facial electromyography demonstrated active right facial neuropathy, and an audiogram demonstrated normal hearing thresholds bilaterally.

Contrast enhanced magnetic resonance imaging (MRI) performed approximately 3 years prior did not reveal any evidence of a facial nerve lesion. However, recent imaging revealed avid enhancement of the right geniculate ganglion with widening of the labyrinthine segment and the geniculate ganglion (►Fig. 1). Computed tomography (CT) demonstrated an expansile lytic process with honeycomb stippling involving the right facial nerve centered on the labyrinthine segment and involving the geniculate ganglion (►Fig. 2).

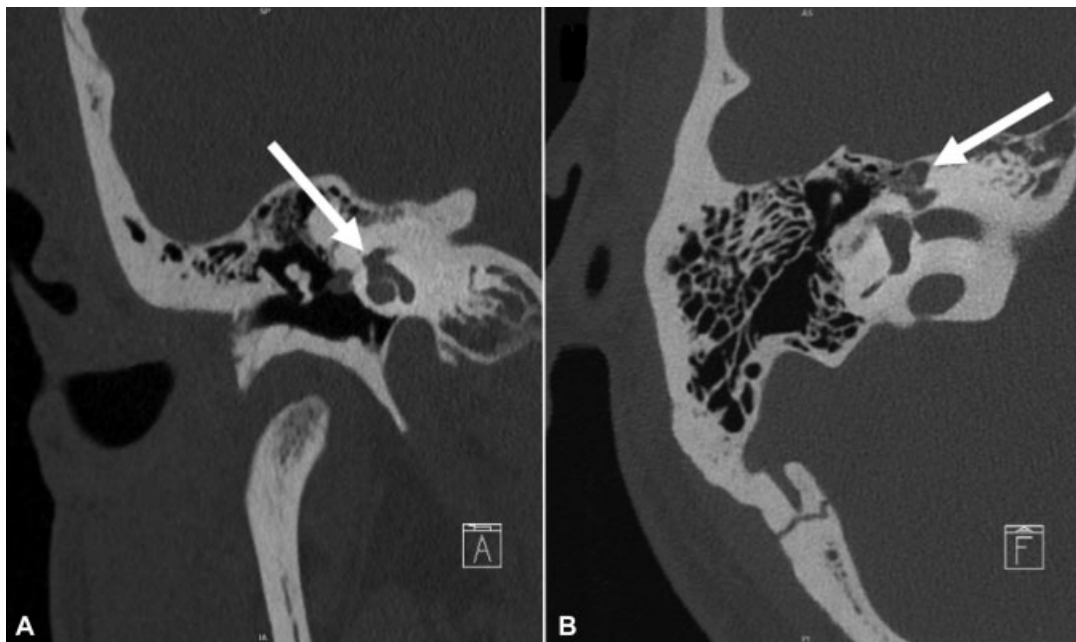


Fig. 2 (A) Coronal and (B) axial temporal bone computed tomography demonstrating expansion of the labyrinthine and geniculate segments of the fallopian canal with honeycomb stippling (intralesional spindle-like calcifications) and a cochlear fistula.

A cochlear fistula was also evident. Radiographic findings were most compatible with a facial nerve hemangioma.

Based on imaging characteristics and poor preoperative facial nerve function, our team offered an option of microsurgical resection with interpositional nerve graft. To access the nerve from the internal auditory canal (IAC) to the mastoid, a combined mastoid-middle fossa craniotomy approach was utilized.

Procedure

A cortical mastoidectomy was performed. The facial nerve was decompressed in the mastoid segment and traced proximally toward the geniculate ganglion. A temporal craniotomy centered over the external auditory canal was used to expose the middle fossa floor. The facial nerve was exposed from the IAC to the geniculate ganglion. A lesion involving the meatal, labyrinthine, geniculate, and tympanic segments was identified and excised. Frozen section analysis revealed atypical cells. Upon removal of the geniculate segment, the cochlear fistula was identified and great care was taken to avoid suctioning of perilymph. A small blood patch was placed over the fistula along with fascia and fibrin glue. A segment of great auricular nerve was obtained through a high cervical incision and was interposed between the meatal and mastoid segments. A neuroorrhaphy tube and fibrin glue were used to secure the graft in place.

Histological sections obtained from formalin-fixed paraffin-embedded tissue demonstrated disorganized nerve fascicles, involving peripheral ganglia. No associated cellular proliferation or abnormal blood vessels were identified (→**Fig. 3**). Immunohistochemical staining with neurofilament (2F11 clone; Dako, Santa Clara, California, United States) highlighted disorganization of the nerve fascicles. In addition, this also demonstrated foci with relative aggregation of axons, suggestive of axonal clusters. There was no proliferation of Schwann cells demonstrated by S100 (polyclonal; Dako) or of perineurial cells revealed by Epithelial membrane antigen (EMA) (E29 clone; Dako). No macrophage aggregates were detected by CD68 (KP1 clone; Dako), and no increase in collagen IV staining (CIV 22 clone; Dako) was seen in association with the disorganized nerve fascicles. Overall,

the appearance of disorganized nerve fascicles, demonstration of axonal aggregates reminiscent of axonal sprouting/regeneration, and absence of any cellular proliferation or vascular malformation were considered to favor a TN. At 3 months postoperatively, MRI demonstrated no evidence of recurrent lesion. Postoperatively, the patient developed ipsilateral profound sensorineural hearing loss.

Discussion

Traumatic facial neuromas are rare entities that comprise a very small fraction of lesions affecting the facial nerve. Prevalence estimates are based on cadaveric temporal bone studies and range between 0.002 and 0.8%.^{1,6,7} As TN of the facial nerve are seldom encountered, more common lesions of the facial nerve must be considered in a patient with facial paralysis and imaging demonstrating an abnormality of the facial nerve along its course. The two most common tumors of the intratemporal or intracranial facial nerve are schwannoma and hemangioma. Neurofibromas of the facial nerve, more common in neurofibromatosis type 1, frequently affect distal branches in the parotid gland but can extend intratemporally.⁸ While malignant peripheral nerve sheath tumors of the facial nerve have been reported, parotid tumors with overt or occult perineural spread account for the most common cause of tumor infiltration causing facial paralysis.⁹ Histopathologically, facial nerve TN demonstrates disorganized nerve fascicles. Demonstration of axonal sprouting with an associated Schwann cell can be best seen by electron microscopic examination of glutaraldehyde-fixed plastic-embedded sections. In this case, given the clinical suspicion of a neoplastic process, only formalin-fixed paraffin-embedded sections were available for evaluation. Even so, the presence of axonal clusters in the disorganized nerve fascicles, highlighted by a neurofilament immunostain, is suggestive of axonal sprouting as part of regeneration. Notably, the absence of a cellular proliferation excludes a neoplastic process.

There have been 14 previously reported cases of TN involving the facial nerve. While blunt trauma without disruption of neural continuity accounts for the most common etiology of

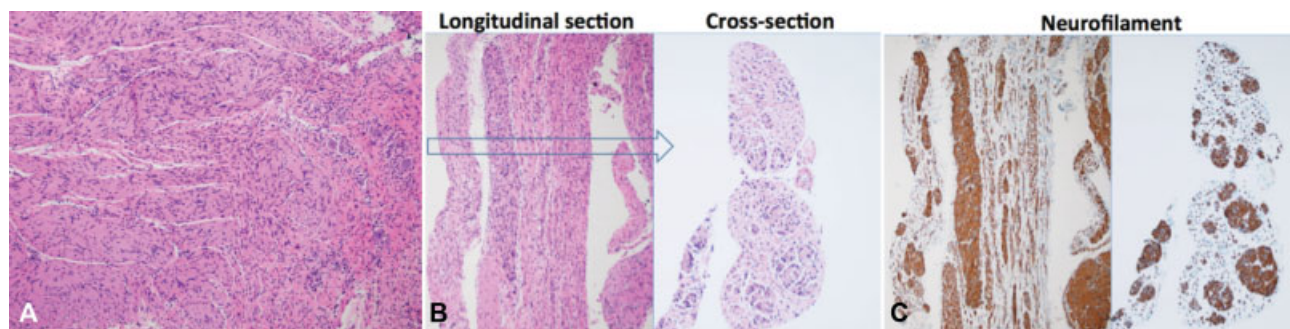


Fig. 3 (A) Histological sections demonstrate disorganized nerve fascicles, which also involved peripheral ganglia (hematoxylin and eosin; $\times 100$ magnification). (B) Longitudinal and cross-sections of the disorganized nerve fascicles, suggesting variably density of the axons (hematoxylin and eosin; $\times 100$ magnification). (C) Neurofilament highlights the axonal disorganization in the longitudinal section and demonstrates axonal aggregates in the cross-sections, suggestive of axonal clustering ($\times 100$ magnification).

facial TN, several cases have been identified following mastoid surgery, thought to be related to middle ear inflammation. Two cases were considered idiopathic.^{3,10,11} In the setting of chronic middle ear or mastoid inflammation, facial nerve function was typically normal and the neuroma was discovered incidentally or on postmortem evaluation.³ In contrast, traumatic facial neuromas are commonly associated with some degree of facial paralysis.^{3,10,11}

Based on location, history, and imaging characteristics, the leading differential diagnosis in this case was geniculate ganglion hemangioma. These “tumors” were originally thought to develop from the rich capillary plexus surrounding the geniculate ganglion. Based on histology and immunohistochemistry, these lesions are now favored to be a venous malformation rather than a true neoplasm. Less commonly, facial nerve hemangioma can occur in the IAC, at the second genu, and near the takeoff of the chorda tympani nerve.¹² In contrast to facial nerve schwannoma, geniculate ganglion hemangioma is classically associated with more advanced nerve symptoms (including spasm, recurrent palsy with intervals of recovery, or progressive facial paralysis) out of proportion to tumor size.¹³ Vertigo and pulsatile tinnitus are rare but may result from invasion of the labyrinth.¹⁴ Facial nerve hemangiomas are typically T1 iso/hypointense, T2 hyperintense, and heterogeneously enhanced on MRI.¹⁴ CT imaging demonstrates ill-defined margins with intralesional bone spicules characterized as a “honeycomb” pattern as seen in the index case.

Given its rarity and overlapping appearance with geniculate ganglion hemangioma and facial nerve schwannoma, definitive preoperative diagnosis is challenging. However, a recent report by Chhabra et al suggest the use of MR neurography as a means of distinguishing TN from neoplasia using features such as perilesional scarring, lack of a split fat or target sign, and absence of abnormal enhancement.¹⁵ In situations with a convincing prior history of trauma, MR neurography may be considered as an adjunct to high-resolution cross-sectional imaging with MR and CT.

While there is no established treatment algorithm for TN, one may adapt treatment strategies for facial nerve schwannoma or geniculate ganglion hemangioma to these rare cases.² The best facial nerve functional outcome yielded by interpositional nerve grafting is HB grade III. A majority of patients enjoy good resting tone, but a portion still require adjunct procedures to aid in eyelid closure. Therefore, if the facial paresis is severe (i.e., HB grades IV–VI), it is reasonable to opt for nerve resection with interposition nerve graft placement or primary reanastomosis. To prevent muscle atrophy and degeneration of motor end plates, it is ideal to proceed with nerve grafting within 12 to 18 months in situations with complete facial paralysis. Unlike in facial nerve schwannoma or geniculate ganglion hemangioma, however, neural preservation strategies (i.e., subtotal resection with observation of residual tumor) are not feasible in TN.

Conclusion

Facial nerve TN is a rare consequence of trauma. Given the overlapping clinical presentation and imaging characteristics of facial TN and other more common primary facial nerve lesions, such as geniculate ganglion hemangioma and facial nerve schwannoma, preoperative diagnosis remains challenging. In general, management of facial paralysis associated with TN parallels that of geniculate ganglion hemangioma and schwannoma—interposition nerve graft or donor nerve procedures should be considered in cases with HB grade IV or poorer function and no evidence of improvement after 6 to 12 months of observation.

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