Aggressive Fibro-Osseous Lesion of the Temporal Bone

Brian Nuyen, BS¹; Christopher G. Tang, MD²; Harold Korol, MD³

¹University of California, San Diego School of Medicine, San Diego, CA
²Department of Head and Neck Surgery, Kaiser Oakland Medical Center, Oakland, CA
³Department of Head and Neck Surgery, Kaiser Permanente Medical Center, Redwood City, CA

ABSTRACT

We present a rare case of a clinically aggressive temporal bone tumor that was described as a benign fibro-osseous lesion (BFOL) on pathology. We also highlight the surgical approach used to resect it and review the literature on fibro-osseous lesions of the temporal bone. This case not only demonstrates a particularly destructive presentation of a temporal bone BFOL, but also emphasizes that temporal bone tumors benign on histopathology may be clinically aggressive.

A 53-year-old male initially presented with a several month history of left-sided hearing loss and non-pulsatile tinnitus. Imaging revealed a destructive lesion eroding through the left temporal bone. The patient received a mastoidectomy followed by a revision mastoidectomy a year later. All pathological specimens were consistent with a benign fibro-osseous lesion. Despite its benign histopathologic appearance, the tumor continued to invade surrounding structures, approaching the jugular bulb. The patient then underwent a resection via a lateral skull base approach, which resulted in dramatic improvement of compressive symptoms. Temporal bone BFOLs are rare and even more rarely feature a relentlessly aggressive clinical course. Early and complete resection seems to be required for such histologically benign yet locally destructive lesions.

INTRODUCTION

Benign fibro-osseous lesions (BFOLs) of bone arise from an insidious, idiopathic process by which normal bone is replaced initially by fibrous connective tissue and then infiltrated by osteoid and cementoid tissue. Variant patterns of ossifications and calcifications with a hypercellular fibroblast marrow element characterize this broad group. The nosology of BFOLs of the craniofacial complex includes bone dysplasias, cemento-osseous dysplasias, inflammatory/reactive processes, metabolic diseases, and neoplastic lesions.¹ While the mandible and maxilla are the most documented craniofacial sites of origin for these lesions, temporal bone involvement is also noted in much smaller case series and isolated case reports.²⁻⁷ In this report, we describe a case of a clinically aggressive temporal bone tumor that was described as a BFOL on pathology. We also highlight how an infratem
poral fossa surgical approach resected this uniquely aggressive, recurrent BFOL in the temporal bone leading to symptom improvement. In reflecting on this case, we also highlight the literature on aggressive fibro-osseous lesions of the temporal bone.

**CASE PRESENTATION**

A 53-year-old Asian-American male initially presented to our head and neck surgery department with a several-month history of left-sided hearing loss and non-pulsatile tinnitus. The patient denied pain, otorhea, and vertigo; furthermore, lower cranial nerve symptoms were not endorsed. The patient had an otherwise non-contributory past medical history, with no known allergies, medications, social/family history of similar symptomatology; review of systems other than head/neck was also normal. Otoscopy was remarkable for an occluded left external auditory canal, with no view of the tympanic membrane. Audiogram revealed mild left-sided conductive hearing loss. Computed tomography imaging and magnetic resonance imaging revealed a 2.3 x 3.1 x 3.3 cm destructive lesion eroding through the left temporal bone towards the jugular foramen (Figure 1A). The lesion had a low T1 and high T2 signal, was mildly heterogeneous in intensity, and enhanced with contrast. Besides opacification of the left mastoid air cells, no other irregularities were observed, including no abnormalities on the right temporal bone region. Despite these findings, the patient opted for observation at the time.

Three years later, the patient underwent a left canal wall-down mastoidectomy with debulking of the primary tumor and meatoplasty. Significant bone erosion with partial erosion of the ossicles as well as a dehiscent facial nerve in the mastoid and tympanic segments was noted intraoperatively. Bulky, benign-appearing pink-tan myxoid tumor tissue was removed piecemeal.

**Figure 1.** Magnetic resonance imaging (MRI) sequences highlighting lesion’s anatomical reach. A. Initial MRI shows a 2.3 x 3.1 x 3.3 cm destructive lesion eroding through the left temporal bone towards the jugular foramen. The lesion had a low T1 and high T2 signal, was mildly heterogeneous in intensity, and enhanced with contrast. No other irregularities were observed on imaging (axial view, with contrast). B. MRI status-post mastoidectomies shows a significant interval increase in size since the initial MRI, with the mass now measuring 4.2 cm in diameter. Mass also noted to extend into the apex of the left internal carotid artery canal with opacification of the skull base at the left internal jugular fossa (axial view, with contrast).
Post-operatively, despite intraoperative visualization of the facial nerve, the patient experienced partial facial nerve paresis (House Brackmann 2/6). Post-operatively, the patient also reported no serviceable hearing on the left, confirmed by audiogram. Patient then received post-operative radiation, which did not affect tumor size.

16 months later, the patient underwent a revision mastoidectomy and posterior fossa craniotomy to remove more tumor and decompress facial nerve. The previously noted dehiscent facial nerve was preserved. The bone fragments that had since regrown around the facial nerve were removed. Furthermore, the nerve’s activity was confirmed by stimulation with nerve monitors. Postoperatively, however, the patient reported no improvement of his partial facial paresis. Despite the second surgery’s excisions, the benign lesion continued to grow over the next year and began causing compressive symptoms on the jugular bulb. MRI obtained at this time showed a significant interval increase in size, with the mass now measuring 4.2 cm in diameter (Figure 1B). The mass was noted to extend into the apex of the left internal carotid artery canal with opacification of the skull base at the left internal jugular fossa.

8 months following the second surgery, the patient received a revision tympanomastoidectomy with a Fisch Type A skull base approach and dissection of jugular foramen and lower cranial nerves. After this surgery, patient’s pre-operative residual facial paresis and profound hearing loss on that side remained; however, his compressive symptoms dramatically improved.

After each surgical extirpation of the tumor, histopathological analysis was performed in consultation with outside institutions. Histologic examination of the mastoid contents and posterior canal wall resected during the patient’s initial mastoidectomy showed fragments of woven bone rimmed by osteoblasts embedded in a fibromyxoid stroma with scattered leukocytes (Figure 2A, 2B, 2D, 2E). The stromal cells were small and bland, with a plasmacytoid appearance in some areas. Immunohistochemical stains showed stromal cells negative for cytokerin, GFAP, CD34, desmin, p63, and beta-catenin; staining was equivocal with S100, SMA, and calponin. Ki-67 proliferation index highlighted less than 10% of the lesional cells (Figure 2C). This all supported the diagnosis of benign fibro-osseous lesion and reduced the possibility of other disease processes, including malignant pathologies.

Histopathology of specimens from the patient’s revision surgery a year later reaffirmed a diagnosis of benign fibro-osseous lesion. The sec-

![Figure 2](https://example.com/fibro-osseous-lesion.png)

**Figure 2.** Resected temporal bone histopathological samples confirming benign fibro-osseous disease. A, B, D, E. High- and low-power hematoxylin & eosin (H&E) stains reveal woven bone fragments rimmed with osteoblasts embedded in a fibromyxoid stroma, with scattered leukocytes. No cytologic atypia or mitotic figures are found (A: 10x magnification; B: 10x magnification; D: 40x magnification; E: 20x magnification). C. Ki-67 stain reveals a ki-67 proliferation index < 10%, reducing the possibility of malignancy (20x magnification). F. S100 stain is negative, with only mast cells showing light, scattered staining (20x magnification).
from the left mastoid specimens obtained from this revision mastoidectomy again showed lamellar and woven bone involved by a pauci-cellular fibromyxoid spindle cell lesion, similar to the previous biopsy. There were associated osteoclast multi-nucleated giant cells, but once again no evidence of cytologic atypia or mitotic figures, and no evidence of more sinister pathologies.

The specimens from the patient’s final surgery showed again bland stromal cells in a fibromyxoid background and were consistent with a benign fibro-osseous lesion. No cytologic atypia or mitotic figures were observed. Furthermore, immunohistochemical stains of these specimens showed scattered staining with S100, possibly mast cells (Figure 2F), and numerous CD68 positive cells that were likely histiocytes. While some of the left mastoid contents showed evidence of cholesteatoma, including numerous anucleate keratinized squamous cells, no malignancy or any other histopathology besides BFOL was identified. Additionally, left Level III lymph nodes obtained during his last surgery were negative for malignancy.

**DISCUSSION**

The differential diagnosis of benign masses of the temporal bone includes skin cysts, cholesteatomas, middle ear adenomas, endolymphatic sac tumors, neuromas/schwannomas, glomus tumors, exostoses, aneurysmal bone cysts, giant cell tumors, eosinophilic granulomas, and BFOLs. Despite a benign histopathological appearance, some of these lesions can be invasive, eroding through temporal bone as well as compressing and invading neighboring structures. Certain BFOLs like fibrous dysplasias (FDs) and ossifying fibromas (OFs) in particular have been noted in case reports and small case series for their expansile characteristics in the temporal bone. FDs are characterized by benign, nonneoplastic, cellular proliferation of fibroblasts, with formation of irregular trabeculae of bone or ovoid calcifications shaped like “Chinese characters” that show indistinct, nonencapsulated borders. The prominent symptom of FD in the temporal bone is progressive hearing loss in roughly 80% of cases, most often conductive type in nature from obliteration of external and middle ear structures. Less common presenting symptoms of FD in the temporal bone include recurrent facial nerve palsy, otorrhea, otalgia, trismus, pulsatile tinnitus, facial numbness, and facial pain. FDs encountered in the operating room are typically unencapsulated without discrete margins; affected bone subtly blends into the normal bone. While some FDs can be observed, bone remodeling and resection are important tools for FD management as there is an approximately 50% recurrence rate with curettage treatment; moreover, complete resection is curative.

OFs are BFOL neoplasms that are confined to the jaws and the craniofacial complex. Histologically, OFs demonstrate a distinctly encapsulated proliferation of cellular fibrous connective tissue with distinct lamellar transformation at the periphery, which contrasts the blurred margins of FD lesions. Predominantly arising from the jaw in 75-89% of cases, OFs less commonly involve the nasal bones, orbit, paranasal sinuses, occiput, and temporal bones. OFs were first described in the temporal bone by Stecker in 1971 and have since been only described in less than ten cases. In contrast, around 200 FDs arising from temporal bone have been described in the literature. On radiograph, OFs appear as well-circumscribed lesions with smooth, often sclerotic borders. OFs are usually unilocular, although multilocularity has been reported, and extent of radiolucency or radiopacity depends on the lesion’s maturity. The well-defined radiological borders correspond with intraoperative findings, as OFs characteristically “shell out” from surrounding bone in intact or large pieces.
secondary to their encapsulations. Clinically, OFs of the temporal bone lead to conductive hearing loss, pain, headaches, and otorrhea as common presenting complaints. While these lesions are typically slow-growing, they may be erosive. Because of this potential for a clinically aggressive course, OF management requires early complete resection with close long-term postoperative follow-up.\(^5\)

In consultation with pathologists from multiple institutions, our case’s histopathological characteristics as described were best characterized as a BFOL. While further specification as OF, FD, or another BFOL subtype could not be definitively ascertained, such subtype designation did not alter the management ultimately required for our patient – complete resection, a management strategy common to clinically-aggressive BFOL subtypes like OF or FD.\(^8\) In contrast to curative resection, radiotherapy proved to be of no benefit in this case. Prior literature has suggested that radiotherapy actually may be associated with sarcomatous degeneration for certain BFOL subtypes, dampening any potential advantage radiotherapy might confer.\(^8\) Furthermore, this lesion stands out in the quality of its expansile nature. While certain temporal bone BFOLs like the OFs and FDs cited above demonstrated significant temporal bone erosion, few other documented temporal bone BFOLs had continued to progress and approach the jugular bulb as aggressively as the lesion of this case, even despite multiple prior resections.\(^2^7\)

Of our patient’s multiple surgeries, the most definitive resection for our patient involved an infratemporal fossa approach. Since first described in 1961 by Fairbanks-Barbosa for advanced tumors of the maxillary sinus, the infratemporal fossa approach to the lateral skull base has since been documented in the successful resection of neurofibromas, giant cell tumors, and glomus tumors in the temporal bone region, as well as trigeminal neuromas and jugular foramen meningiomas.\(^9^12\) With its ability to access the jugular foramen, petrous apex, clivus, and the parasellar and paraphenoidal compartments, the infratemporal approach complements the middle fossa and posterior (translabyrinthine) approaches in the surgical management of most hard-to-reach lesions in the temporal bone. While past clinically aggressive BFOLs in the temporal bone have been removed via transmastoid and combined transmastoid/middle fossa approaches,\(^5\) the infratemporal approach proved ideal to resect our patient’s mass, especially in its extensions close to the jugular foramen. Besides completing the repertoire of surgical access to some of the most hard-to-reach areas of the lateral skull base, it is important to note that the infratemporal approach notably achieves preservation of the inner ear function in the standard hearing patient, avoiding the mandatory labyrinthectomy demanded by a translabyrinthine approach. The outcome of the BFOL resection in this case appeared favorable in our patient’s case, but larger case series are needed for a more conclusive assessment.

The infratemporal fossa approach warrants awareness of certain complications. Cranial nerve polyneuropathy is a prominent risk, with sequelae like corneal abrasion, neurotrophic ulcers, and jaw asymmetry. Rarer complications include wound necrosis, infection, and cerebrospinal fluid leak.\(^13\) The outcome and prognosis of this infratemporal approach for masses in this region of the lateral skull base are dependent on the completeness of resection and the histological type of the tumor.

LEARNING POINTS

\begin{itemize}
  \item There may be a disconnect between clinical course and histological assessment as tumors which appear benign on
histopathology may clinically behave aggressively.

- The infratemporal fossa approach should be considered as a key tool in the surgeon’s armamentarium for hard-to-reach lesions of the lateral skull base.
- Early and complete resection seems to be required for such histologically benign yet locally destructive lesions.

ACKNOWLEDGEMENTS

The authors would like to thank Stanford Pathology and Kaiser Permanente Redwood City Pathology for their consultative input on this case.

REFERENCES


