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CASE REPORT

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Pediatric intraosseous cranial myxoma: A case report and review of literature

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Abstract

While intraosseous cranial myxoma is a rare pathology, it is important for providers to be aware of it, as early diagnosis and treatment is imperative for prognosis. Long-term follow-up is needed as high rates of recurrence have been documented.

KEYWORDS

intraosseous myxoma, occipital skull myxoma

1 | INTRODUCTION

A myxoma is rare benign tumor of mesenchymal origin. In terms of head and neck myxomas, primary tumors are rare and most often arise from the mandible, maxilla, and oral cavity.[1,2] Primary myxomas outside of this region, such as cardiac myxomas, can metastasize to the head and neck.[1] Myxomas can occur at any age but have the highest incidence between 20–30 years old. No gender predilection has been noted.[2,3]

There is a paucity of data on incidence and treatment of intraosseous occipital myxomas. Case reports have identified occurrences of other intracranial myxomas. In this case report, a 12-year-old boy with a myxoma arising from the occipital bone is presented due to the tumor's rare anatomical location, especially in a pediatric patient.

2 | CASE REPORT

A 12-year-old boy presented to the Emergency Department with left ear pain with tenderness to palpation overlying the left mastoid bone. The history revealed that the pain had been present intermittently 1 year but had acutely worsened prior to presentation. There were no associated otologic symptoms including hearing loss, tinnitus, otalgia, otorrhea, vertigo, or facial weakness. A computed tomography scan (CT) with contrast at this time showed

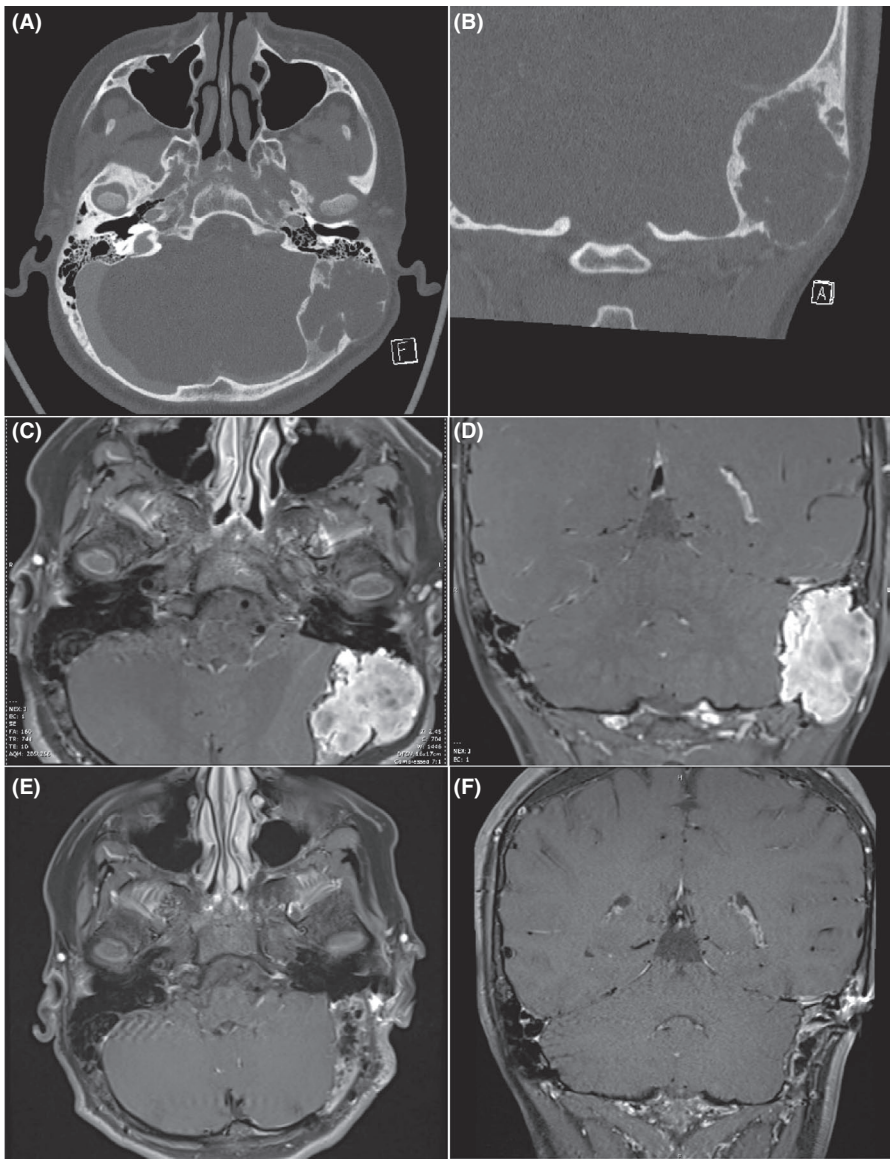
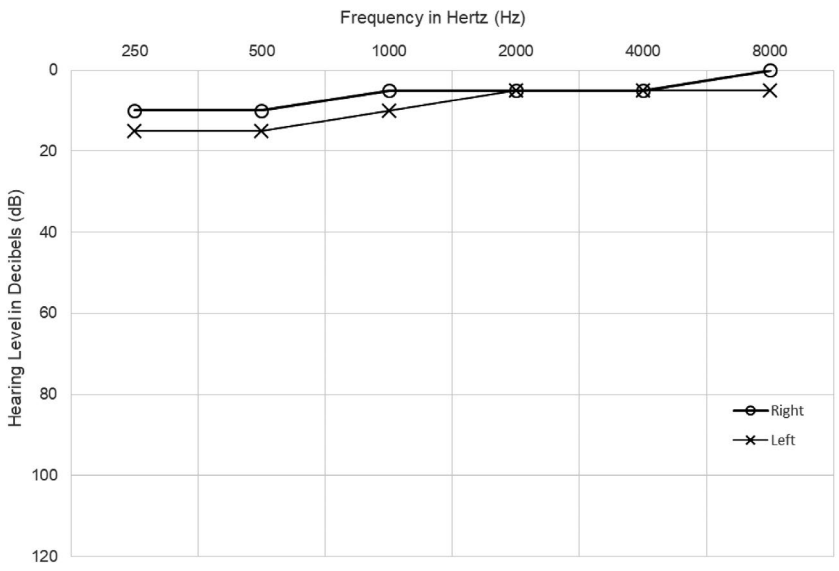


FIGURE 1 (A-B) Axial (A) and coronal (B) post-contrast computed tomography (CT) scan of the left temporal bone lesion on presentation; (C-D) Axial (C) and coronal (D) post-contrast T1 magnetic resonance imaging (MRI) of the lesion on presentation; (E-F) Axial (E) and coronal (F) post-contrast T1 magnetic resonance imaging (MRI) of the surgical site 10 months postoperatively showing fat graft in place and no evidence of recurrent tumor



	Right	Left
Speech Reception Threshold to Spondees (dB)	10	10

FIGURE 2 Audiogram showing hearing within normal limits bilaterally

a 4.8 cm lytic mass centered in the occipital bone bordering the mastoid. Contrasted magnetic resonance imaging (MRI) demonstrated an enhancing mass with measurements of 4.4 × 3.5 × 4.6 cm (Figure 1A-D) and associated mass effect in the left cerebellar hemisphere. Venogram demonstrated non-visualization of the left sigmoid sinus. Audiogram showed bilaterally normal hearing (Figure 2).

The patient underwent biopsy of the suboccipital mass. A tan-colored jelly-like intraosseous tumor was encountered (Figure 3). Initial pathological workup was inconclusive but thought to be suspicious for low-grade sarcoma. Definitive resection with left occipital craniotomy and gross total resection was performed 1 month later through a post-auricular incision. Intraoperative findings were consistent with erosion through bone without dural involvement. Tumor was removed from the mastoid tip and occipital bone under the operating microscope. Areas of dura were exposed, but no CSF leak was encountered.

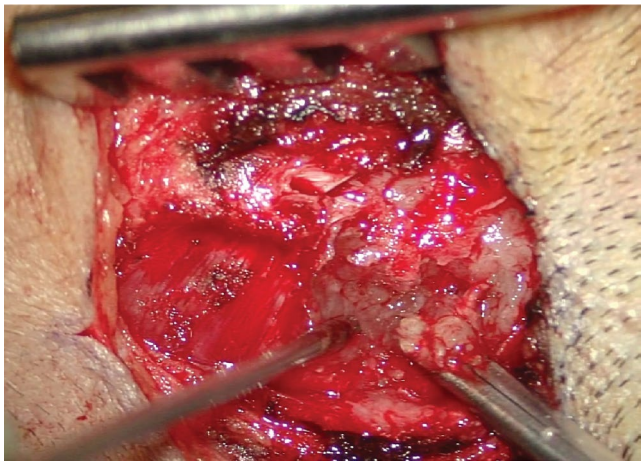


FIGURE 3 Intraoperative photograph from initial biopsy showing gross appearance of the gelatinous tan-colored tumor

An abdominal fat graft and Medpor (Stryker corporation, Kalamazoo, MI) implant were utilized to reconstruct the defect in the occipital bone.

Histology of the final specimen demonstrated loose amphophilic lobules composed of interconnecting pink septae of collagenous fibrous tissue. These septae were surrounded by hypocellular aggregates composed of cytologically bland spindle and stellate cells appearing to be fibroblasts in nature as stained by vimentin only. These cells were enmeshed in a basophilic stroma that contained branching capillaries and occasional macrophages and lymphocytes. Immunohistochemistry was positive for vimentin and focally positive for actin (Figure 4A-D). It was negative for all other markers including CD34, CD1a, desmin, myogenin, MyoD1, keratin cocktail, EMA, S100, SOX10, HMB45, GFAP, MUC4, MDM2, ERG, GLUT1, keratin AE1/3, beta catenin, CD31, FLI1, and CD68. These findings were most consistent with an intraosseous myxoma.

Follow-up with MRI at 10 months showed a stable fat graft with no persistent or recurrent tumor (Figure 1E-F). No postoperative complications, neurological deficits, hearing changes, or evidence of recurrence were apparent on clinical exam. The patient has intact facial nerve function and no noticeable cosmetic defect.

2.1 | Literature review

Including the case presented here, eleven reports of cranial myxomas were found in the literature. Age at presentation ranged from 11–56 years old with an average of 30.4 years and a 7:5 male:female distribution. Presentation varied depending on site of origin, but common symptoms included hearing loss, aural fullness, and visual impairment. The average size on initial imaging across longest

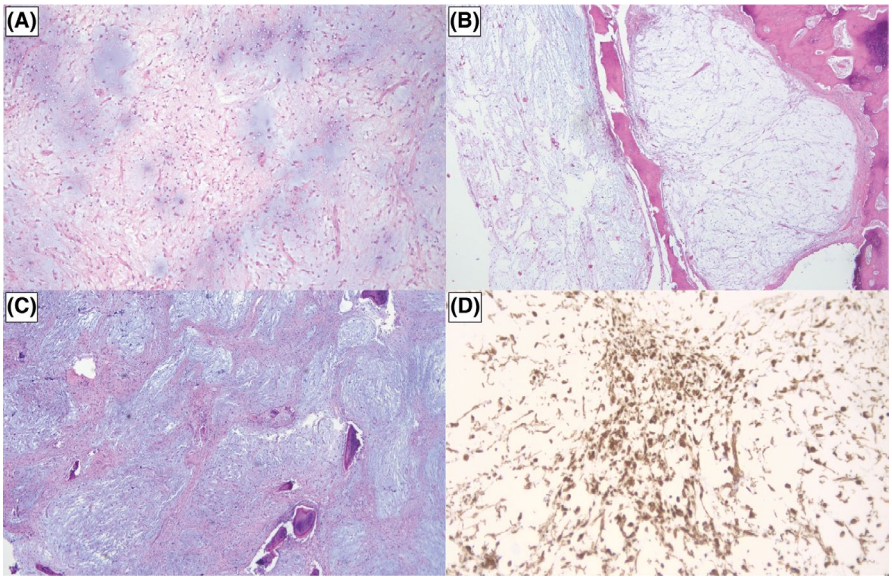


FIGURE 4 (A) Lobular architecture composed of interconnecting septae of collagenous fibrous tissue surrounding aggregates of hypocellular tumor enmeshed in a prominent flocculent basophilic stroma. (B) Tumor infiltrates preexisting cancellous bone. (C) Tumor consists of cytologically bland spindle and stellate cells. (D) Immunohistochemical stain for vimentin is positive in tumor cells

TABLE 1 Literature review of primary skull-base myxomas

Authors	Age (years), sex	Symptoms	Location of tumor	Size (cm)	Intra-osseous?	Immunostain	Treatment	Compli-cations	Prognosis
Oruckaptan et al.[1]	34, M	Right facial weakness	R. temporal bone, middle fossa	6.4 × 4.8 × 5.7 cm	Yes	(+) vimentin, (−) pan-cytokeratin, keratin, S−100	Total resection	None	Local recurrence at 3-year follow-up
Osterdock et al.[5]	17, M	Left sided hearing loss and headaches	L. temporal bone, petrous	n/a	Yes	(−) S−100	Total resection	None	n/a
Nagatani et al.[6]	38, M	Visual impairment	Posterior fossa	n/a	No	n/a	Total resection	None	No recurrence at 4-year follow-up
Ulku et al.[3]	42, F	Right sided hearing loss and fullness	R. middle ear	n/a	Yes	(+) CD34 (−) S-100	Total resection	None	No recurrence at 36-month follow-up
Yin et al.[4]	27, M	Visual impairment and epistaxis	R. temporal bone, ethmoid	8.5 × 8.0 × 5.8 cm	Yes	(+) vimentin, (−) CD34, pan-cytokeratin, keratin, and S-100	Total resection	None	No recurrence at 6-month follow-up
Hasnaoui et al.[2]	53, F	Right sided hearing loss and tinnitus	R. external auditory canal	2.0 × 1.0 cm	Yes	n/a	Total resection	None	No recurrence at 1-year follow-up
Ryu et al.[7]	50, M	Vertigo and hyposmia	R. ethmoid sinus	8.0 × 6.7 × 5.4 cm	No	(+) vimentin, (−) GFAP, S-100, EMA, CD34, and cytokeratin	Total resection	None	No recurrence at 3-month follow-up
Sareen et al.[8]	11, F	Chronic suppurative otitis media	L. temporal bone	2.0 × 1.0 cm	Yes	n/a	Total resection	None	No recurrence at 2-year follow-up
Srinivasan[9]	14, F	Right sided otalgia, diplopia	R. petrous bone	2.9 × 2.5 × 2.3 cm	Yes	(−) S-100	Sub-total resection	None	No recurrence at 1-year follow-up
Ito et al.[10]	11, F	L. homonymous hemianopia, papilledema	R. temporal bone	n/a	Yes	n/a	Total resection	None	No recurrence unknown follow-up timeline
Charabi et al.[11]	56, M	L. chronic otitis media	L. temporal bone	n/a	Yes	n/a	Total resection	Contin-ed otitis media	n/a
Howser et al.	12, M	Left sided otalgia	L. occipital bone	4.4 × 3.5 × 4.6 cm	Yes	(+) vimentin, (−) CD34, keratin, EMA, GFAP, and S-100	Total resection	None	No recurrence at 3-month follow-up

axis when reported was 4.9 cm. Pertinent histopathological features include vimentin positivity as well as cytokeratin and S-100 negativity. These features were consistently observed across the tumors reported. All patients except one underwent total resection. No significant complications were reported. Mean follow-up was 20 months. Only one patient was reported to have a local recurrence at 3-year follow-up. (Table 1).

3 | DISCUSSION

Although the typical location for a myxoma is the cardiac atrium, the tumor may rarely present from bone and other soft tissues. Primitive embryonic mesenchyme is thought to be the possible origin of skull-based myxomas, which is present in the mastoid, ethmoid, and sphenoid air cells of embryos.[4] These tumors are benign but can be locally aggressive. There is a high incidence of recurrence between 3 months and 10 years after surgical resection.[3,5]

An intraosseous myxoma can be identified by degree of gross invasion, histopathology, and immunochemical staining. An intraosseous tumor must invade the bone. This can be identified on imaging or direct visualization in surgery. Histopathology can help identify the classic features of intraosseous myxoma tissue, such as bland spindle cells and myxoid cytoplasm. Immunochemistry stains positive for vimentin, an intermediate filament present in non-muscle mesenchymal cells.

The differential diagnosis of this case included low-grade myxoid fibrosarcoma, osteo-chondromyxoma, ossifying plexiform tumor, ossifying fibromyxoid tumor, cystic fibrous dysplasia, Langerhans cell histiocytosis, or metastatic disease. However, none of these had sufficient clinical evidence or immunohistochemistry to support a diagnosis. The lack of diffusion argued against hypercellular tumors such as rhabdomyosarcoma or metastatic neuroblastoma. While occipital bone myxoma is a rare diagnosis, it is nonetheless one that providers should be aware of since the recommended treatment differs from other similarly presenting tumors.

The treatment for myxomas primarily includes radical surgical removal of the tumor when possible. Myxomas are thought to be insensitive to radiation therapy, though this may be used as a salvage treatment if radical resection is impossible.[5] Most importantly, timely recognition and early intervention are the best avenues to prevent additional long-term morbidity. The diagnosis of an intraosseous myxoma is not typically at the forefront of the differential, especially in a pediatric patient. Therefore, this case should provide an example of low morbidity and nonrecurrence with multidisciplinary collaboration and early intervention.

4 | CONCLUSION

The suboccipital region is a rare location for an intraosseous myxoma to occur, especially in a pediatric patient. Early diagnosis and treatment are imperative for prognosis, as radical resection is the best chance for nonrecurrence. Even with surgical resection, long-term follow-up is recommended due to high rates of recurrence.

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CONFLICT OF INTEREST

The authors have no conflict of interests to disclose. This work has not been submitted for publication elsewhere.

AUTHOR CONTRIBUTIONS

Lauren A. Howser, BS—IRB application, chart review, literature review, manuscript writing, final approval. Michael J. Ye, MD: Chart review, literature review, manuscript writing, final approval, multidisciplinary coordination, corresponding author. Sampson Boham, DO: Provided pathological images and interpretation, chart review, literature review, manuscript writing, final approval. Rong Fan, MD: Provided pathological images and interpretation, chart review, literature review, manuscript writing, final approval. Rick F. Nelson, MD, PhD: Chart review, literature review, manuscript writing, final approval, provided patient, senior author.

ETHICAL STATEMENT

This manuscript is the author's original work. This manuscript has not been previously published elsewhere and is not under consideration for publication elsewhere. This paper reflects the author's own work in a truthful and complete manner. All authors have been personally and actively involved in substantial work leading to the paper and will take public responsibility for its content.

CONSENT

I confirm that written patient consent has been signed and collected in accordance with the journal's patient consent policy.

INSTITUTIONAL REVIEW BOARD

Exempted by the Indiana University Institutional Review Board [Protocol ID #11254].

DATA AVAILABILITY STATEMENT

The data used to create this manuscript are available upon request to the corresponding author.

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