Juvenile Ossifying Fibroma: Successful Endoscopic Gross Total Resection of a Rare Sinonasal Tumor in an Adolescent Male

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Abstract

Objectives: To report a rare case of Psammomatoid Juvenile Ossifying Fibroma with extension into the orbit and anterior cranial fossa successfully removed by an endonasal endoscopic approach in an adolescent patient.

Study Design: Case report with literature review.

Methods: PubMed search and case report.

Results: The endonasal endoscopic approach was successful in obtaining a gross total resection of this large tumor that extended lateral into the orbit and superior into the anterior cranial fossa across the midline. Endoscopic office evaluation and followup CT scan is consistent with no evidence of recurrent disease at more than six months.

Conclusions: Juvenile ossifying fibroma is a rare tumor of the sinonasal cavity and more unusual in the pediatric age group. Large tumors involving the orbit and cranial fossa have traditionally been resected using an open transfacial/transcranial approach with an open transcranial and endonasal endoscopic approach for smaller tumors. We describe a case in which visualization with straight and angled telescopes and endoscopic instrumentation allowed high confidence resulting in a gross total resection of this very large tumor. We believe this approach by an experienced endoscopist can offer equal success in treatment outcome with lower morbidity and quicker recovery than the traditional approaches for this tumor.

Introduction

Ossifying fibromas, first described in 1872 by Menzel, are rare benign lesions usually found in the tibia and fibula in children. These tumors have also been described in the head and neck region. Most are found in the mandible (62-89%) but the maxilla, orbit, the skull base, and the calvarium have also been reported. While Ossifying Fibroma in the sinonasal tract tends to occur in older patients, (third and fourth decades), Juvenile Ossifying Fibroma (JOF) of the head and neck tends to occur in a much younger population. It can be found in one or multiple sinuses. Presenting symptoms often are related to the mass effect depending on the size and location of the lesion and include pain, pressure, changes in vision, and headache.

JOF, although benign, is a locally aggressive tumor. Typically, a combined transcranial and transfacial approach for removal and repair has been used especially for very large tumors. With the advent of angled telescopes and more advanced equipment to successfully remove large skull base tumors, extended endoscopic approaches have become increasingly popular. We present an adolescent male with a large Juvenile Ossifying Fibroma of the nasal cavity extending into the anterior cranial fossa. A transnasal endoscopic approach allowed gross total resection of this tumor. The child remains disease free 13 months since his resection.

Case Report

14-year-old white male was referred to the Pediatric Otolaryngology clinic for evaluation of nasal congestion worse on the right for several years. Previous skin testing by the allergy clinic revealed positive response to trees, grasses, weeds, and molds. Fluticasone and Olopatadine were used with no improvement in symptoms. Otolaryngologic evaluation revealed an adolescent male with right mild proptosis. Nasal endoscopy identified a mass in the right nasal cavity. CT scan with contrast was performed (figure 1) followed by an MRI to further delineate the extent of the intracranial disease. Operative nasal endoscopy with biopsy provided the diagnosis of Psammomatoid Juvenile Ossifying Fibroma (figure 2). The child returned to the operative theater three weeks later for definitive surgical extirpation with a combined neurosurgical and otolaryngologic team. Pre-operative embolization was performed 24 hours prior to the resection. Total nasal removal of the tumor included middle turbinates resection, total ethmoidectomy and orbital decompression, maxillary antrostomy, sphenoidotomy, frontal sinostomy and removal of the tumor from the anterior cranial fossa. The defect was successfully closed with a nasal-septal flap based on the sphenopalatine artery. The child was admitted to the neurosurgical service and remained in the Pediatric ICU. He was discharged on post-operative day 7. Follow up CT scan at six months postoperatively revealed no evidence of recurrence (figure 3). He remains more than one year disease free.

Discussion

• Ossifying Fibromas (OF) are rare benign lesions of the craniofacial skeleton that replace the normal bone of the face with fibrous cellular stroma.
• Juvenile Ossifying Fibromas (JOF), a variant of this disease, usually occurs in patients in the second decade of life or before and has a slight male predominance.
• JOF is split into two different groups: Juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF).
• JPOF usually occurs in the orbit and paranasal sinuses but has also been found in the mandible, maxilla, and calvarium.
• JTOF is usually found in the maxilla.
• Histology confirms the diagnosis. JPOF shows small masses of uniform calcified osteoid bodies found in a fibroblastic stroma. Osteoclastic giant cells are common. JPOF differs from JTOF as the later has areas of fibrillar osteoid and woven bone with no psammoma like structures.
• Symptoms of JPOF are mainly caused by mass effect and include pain, facial pressure, displacement of the orbit, proptosis, nasal obstruction, and facial swelling.
• Total surgical resection at the earliest stage is the treatment of choice in patients with JPOF. Radiation is not indicated and may increase malignant transformation.
• Smaller tumors are usually resected by an endoscopic approach.
• Larger tumors that involve multiple sinuses are usually approached in an open manner, usually through a transfacial, transcranial or combined approach.
• Recurrence of JPOF has been reported as high as 30-56%. This is likely due to tumor that is inadequately removed. Close follow-up with endoscopic exams and follow-up imaging, to evaluate early recurrence is essential.
• This case outlines a completely endoscopic approach to surgical extirpation of a large tumor with intracranial and orbital involvement.

Conclusion

Juvenile psammomatous ossifying fibroma (JPOF) is a rare disease that occurs mainly in the sinonasal tract in the adolescent patient. These neoplasms have been historically removed both endoscopically (mainly for small tumors) and through open approaches for larger tumors or those extending into the cranial cavity. This paper outlines a case in which a large tumor was successfully removed entirely by an endoscopic approach with high confidence of total gross resection. These patients must be followed closely as there is a high risk of recurrence. We believe this to be the first reported case of such a large tumor in a pediatric patient removed completely through an endonasal endoscopic approach.

References