Case Report

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

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Abstract

Juvenile ossifying fibroma is a rare tumor of the sinonasal cavity in pediatric patients. Large tumors involving the orbit and cranial fossa have traditionally been resected using an open craniofacial 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 open procedures for this tumor.

ABBREVIATIONS

JOF: Juvenile Ossifying Fibroma; OF: Ossifying Fibromas; JPOF: Juvenile Psammomatous Ossifying Fibroma; JTOF: Juvenile Trabecular Ossifying Fibroma

INTRODUCTION

Ossifying fibromas, first described in 1872 by Menzel [1], are rare benign lesions commonly found in the tibia and fibula in children. However, these tumors have also been described in the head and neck region. Most are found in the mandible (62-89%) but lesions in the maxilla, the orbit, the skull base, and the calvarium have also been reported [1].

Ossifying fibromas in the sinonasal tract tend to occur in older patients in the third or fourth decades and are more prevalent in women than men [1]. They are usually characterized as slow growing painless tumors. In contrast, Juvenile Ossifying Fibroma (JOF) occurs in a much younger patient population. Although benign, these tumors are locally aggressive and can cause bony and soft tissue destruction and impingement on the surrounding organs such as the eyes and brain. Presenting symptoms include those related to mass effect, mainly pain, pressure, changes in vision, and headache depending on the size and location of the lesion.

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 superiorly into the anterior cranial fossa, and lateral into the orbit. A transnasal endoscopic approach allowed gross total resection of this tumor. Reconstruction with a nasal-septal flap was successful to close the skull base defect. We believe this to be the first case of such a large tumor in a pediatric patient successfully removed endoscopically. The child remains disease free 18 months since his resection.

CASE PRESENTATION

A 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 response to trees, grasses, weeds, and molds. Fluticasone and Olopatadine nasal sprays were used with no response for six months. Otolaryngology evaluation on presentation noted mild right 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 Juvenile Ossifying Fibroma (Figure 2). The child returned to the operative theater three weeks later for definitive surgical extirpation. Endoscopic endonasal resection of the tumor included ipsilateral middle turbinate resection, total ethmoidectomy, orbital decompression, maxillary antrostomy, sphenoidotomy, and frontal sinusotomy, all together allowing appropriate instrument access for removal of the tumor from the anterior cranial fossa. The defect was successfully closed with a nasal-septal flap based on the posterior septonasal branch of the sphenopalatine artery. The child was admitted to the neurosurgical service in the Pediatric ICU. He was discharged on Post-operative day seven. The child remains disease free for eighteen months as followed by office nasal endoscopy and follow up CT scan (Figure 3).

**SURGICAL PROCEDURE**

Preoperative embolization was performed 24 hours prior to the surgical procedure by the Interventional Neuroradiology Division. The following day the child was brought to the operating theater and general anesthesia was provided. The Medtronic Fusion® image guidance system was registered and used throughout the procedure to verify the tumor confines. Endonasal endoscopic approach was performed using straight and angled rigid telescopes. A large portion of the tumor was within the nasal cavity, with mass effect into the ipsilateral paranasal sinuses. The tumor surface was smooth and mucosalized. It extended into the anterior cranial fossa medially and superiorly, and also extended laterally displacing the right orbit.

The intranasal component of the tumor was removed with the Arthrocare® Coblation device with a sino-nasal handpiece (Procise EZ View Plasma wand ‘EICA 8875-01) set on coagulation 9, and ablation 5 (Figure 4). This allowed for tumor debulking while maintaining adequate hemostasis.

Full access to the tumor confines for visualization and instrumentation required sinusotomies and resection of the middle and superior turbinates. The uncinate process was identified and removed and a large maxillary antrostomy was performed. A total ethmoidectomy was performed to allow both medial and lateral tumor access. The lateral aspect of the tumor displaced the medial orbital wall laterally. Tumor was removed laterally to include removal of the lamina papyracea while leaving the periorbita intact. This cleared the lateral extent of the tumor margin. Posteriorly, a large sphenoidotomy was required to clear...
access to the posterior tumor margin. Superiorly, the frontal sinus ostium was identified and enlarged. The superior aspect of the tumor extended into the anterior cranial fossa and was removed off the dura with neurosurgical curettes and dissectors (Figure 5). Different angled telescopes allowed visualization during dissection and removal of this tumor that extended across the midline in the anterior cranial fossa, along the olfactory cleft and the cribiform plate. The superior dome cortex of the tumor formed a thin shell, being the superior margin of the tumor, which was fairly adherent to the overlying dural. This required meticulous dissection in order to minimize the extent of dural defect. Several small durotomies were required to remove the entire adherent tumor, assuring a gross total resection (Figure 6). The final dural defect was less than one centimeter in greatest dimension, and was repaired using a single layer onlay reconstruction using a vascularized naso-septal flap based on the posterior nasoseptal vascular bundle of the sphenopalatine artery (Figure 7).

DISCUSSION

Ossifying Fibromas (OF) are rare benign lesions of the craniofacial skeleton, usually found in patients twenty to thirty years old. They appear to affect women more than men. The cause of these tumors is currently unknown however theories include derivatives of periodontal ligaments, trauma, or developmental causes [1]. These neoplasms replace the normal bone of the face with fibrous cellular stroma that usually presents as a large mass with deficits related to mass effect on surrounding structures.

Juvenile Ossifying Fibromas (JOF), a variant of this disease, occurs in patients of a much younger age [2]. JOF is split into two different groups: Juvenile Psammomatous Ossifying Fibroma (JPOF) and Juvenile Trabecular Ossifying Fibroma (JTOF) [2]. Differential diagnosis includes fibrous dysplasia, sinonasal Psammomatous Meningioma, and osteosarcoma [1]. The patient in this case was found to have JPOF which will be the focus of this discussion.

JPOF was first described by Benjamins in 1938 [3]. Most are diagnosed before the age of 15 and has a slight male predominance [3]. JPOF usually occurs in the orbit and paranasal sinuses but has also been found in the mandible, maxilla, and calvarium. JTOF, on the other hand, is usually found on the maxilla or mandible. Symptoms of JPOF are mainly caused by mass effect and include facial pain, facial pressure, sinusitis, nasal congestion, displacement of the orbit, blurry vision, headaches, proptosis, and facial swelling [2]. The lesions are slow growing but can be locally aggressive and destructive.

On imaging, JPOF is a well-circumscribed lesion with varying degrees of radiolucency and radiodensity [2]. These lesions are usually very well demarcated from the surrounding bone with a lytic border [2]. Depending on the size and the aggressiveness of the lesion, bony destruction can be found and extension into the intracranial space, although rare, has been described. On MRI, the lesions are hypointense on T1 and T2, however they enhance with gadolinium [4].

Histology of the JPOF show small masses of uniform calcified osteoid bodies that classified as psammomatoid found in a fibroblastic stroma with spindle cells [5]. These psammomatoid particles have a central basophilic area with an eosinophilic border. Osteoclastic giant cells are also a common finding in these tumors [6]. Aneurysmal bone cysts are a common feature of JPOF and can distinguish these tumors from other types of fibrous anomalies [5]. JTOF, for instance, is also composed of fibroblastic spindle cell stroma with multiple multinucleated giant cells. Unlike JPOF, however, cystic structures are rare in this type of lesion [7].

Total surgical resection at the earliest stage is the treatment of choice in patients with JPOF. This can be done in an endoscopic
or an open approach. Smaller tumors are usually easily resected by an endoscopic approach. Larger tumors, however, that involve multiple sinuses are commonly approached in an open manner. The extent and location of the tumor will determine the approach in these tumors.

Transfacial or cranial approaches have been used for open resection. These include the transfrontal nasal approach and the transfrontal nasoorbital approaches [8]. Advantages of open resection include large exposure of the lesion for careful dissection [9]. These methods, however, are extensive surgeries that can cause external scarring, long hospital stays and recovery times for young patients. They require coordination amongst multiple surgeons and disciplines including Otolaryngologists and Neurosurgeons. Endoscopic approaches have been described in small tumors but not in tumors involving the anterior cranial fossa and orbit. This case demonstrates the ability to resect this tumor with careful dissection using endoscopic methods only.

Recurrence of JPOF has been reported as high as 30-56% [8] and this is likely due to tumor that is inadequately removed. Intracranial extension is rare but few cases are reported. These lesions may require larger resections [7]. After total resection, the patient must have close follow up with endoscopic and imaging exams to evaluate for recurrence. Radiation therapy is not indicated in juvenile ossifying fibromas as it may increase malignant transformation from 0.4% to 40% [1].

Juvenile ossifying fibroma is a rare tumor of the sinonasal cavity and even more unusual in the pediatric age group. Large tumors involving the orbit and cranial fossa have traditionally been resected using an open transnasal/ 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. Our case report appears to be the first report of such a large tumor with intracranial and orbital involvement in a pediatric patient removed completely through a transnasal endoscopic approach. The child remains disease free after eighteen months.

REFERENCES