



A Case of Ossifying Fibroma of the Skull Base Misdiagnosed as Hemangioma

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Abstract

Ossifying fibroma is a rare disease. This disease is often found incidentally and may be asymptomatic, but it can also present with symptoms such as headache. Surgical treatment is necessary to alleviate the patient's symptoms. It is important to differentiate it from other diseases such as fibrous dysplasia, cementoblastoma. The authors report a successful surgical treatment case of ossifying fibroma of the skull base misdiagnosed as hemangioma and describes its treatment.

Keywords: Ossifying fibroma; Headache; Hemangioma; Treatment

Introduction

Ossifying fibroma is a rare disease, characterized by a benign tumor with well-defined boundaries. It is usually asymptomatic but is often discovered incidentally when patients complain of headache and other symptoms. Surgical treatment is required to relieve the patient's symptoms and it is important to be differentiated from fibrous dysplasia and cementoblastoma. The authors report a case of ossifying fibroma of the skull base that was initially mistaken for a hemangioma.

Case Presentation

A 40-year-old female patient presented to the outpatient department with symptom of headache. The headache had been present for the past 2 to 3 months and were not present before that. The patient did not complain of any other unusual symptoms. There were no underlying medical conditions such as hypertension or diabetes, and no history of surgery or facial trauma. In the outpatient department, The physical examination, endoscopy and computed tomography were performed. The physical examination and endoscopy revealed no abnormal findings, except for a deviated nasal septum on the left side. The computed tomography scan revealed a 20 mm osteophytic mass lesion in the ethmoid sinus area of the left nasal cavity. The author planned endoscopic surgical treatment. Magnetic resonance imaging was performed for further diagnostic differentiation due to the presence of soft tissue features within the osteoid mass seen on the computed tomography scan (Figure 1). Following magnetic resonance imaging, the patient was referred to the radiology department for further discussion of the lesion diagnosis. The radiology staff diagnosed the lesion as a hemangioma. Based on this diagnosis, additional preoperative procedure (mass feeding vessel embolization) were planned. However, during the neurosurgical evaluation procedure, no feeding vessels were observed within the lesion, and embolization was not performed. Therefore, the author proceeded with endoscopic endonasal mass removal without embolization. The patient's surgery was performed by a single author (only one doctor) and a single assistance nurse, with intraoperative assistance provided by the nursing team. The lesion was successfully removed using endoscopic instruments, such as an endoscopic drill (Figure 2). The exterior of the lesion was hard, but the interior of the lesion appeared soft with osteophytic soft tissue. No hemorrhagic tissue was observed inside or outside the lesion. Although there was no skull base defect observed endoscopically, the lesion was treated postoperatively with TachoComb® (Fibrinogen/thrombin-based collagen fleece, Nycomed, Linz, Austria), Surgicel® (Oxidised cellulose, Ethicon, North Ryde, NSW, Australia) and Floseal® (Hemostatic sealant, FS, Baxter Healthcare, Deerfield, IL, USA) to reinforce any possible skull base defect during surgery. The patient was followed-up in the outpatient clinic as scheduled. The patient's histopathology was diagnosed as ossifying fibroma (Figure 3). Until the latest follow-up (seven months after surgery), the headache, which was the main symptom before surgery, has

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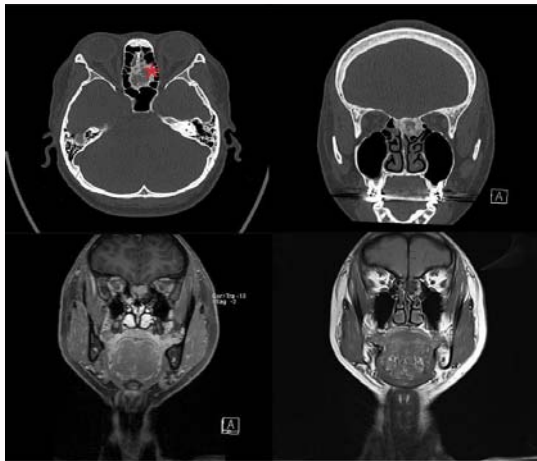


Figure 1: Pre-operative evaluation. Computed tomography axial and coronal plane images (above two figures of entire figures) and magnetic resonance imaging T1 and T2 coronal plane images (below two figures of entire figures). The asterisk presents the lesion of ethmoid sinus adjacent to skull base.

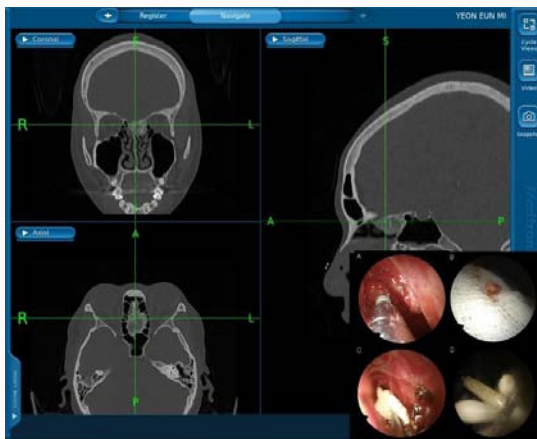


Figure 2: Screenshot image of navigation-guided transnasal endoscopic finding. The point of green cross-line shows the lesion of ethmoid sinus intraoperatively. Mini-square (A-D) pictures present the endoscopic finding during the operation. A) The endoscopic diamond drill bur was used to remove ossifying fibroma bony lesion; B) The ossifying fibroma biopsy was obtained; C, D) Hemostatic materials (Surgical® and Floseal®) were inserted to the removed ossifying fibroma lesion to perform postoperative bleeding control.

disappeared. The olfactory function has recovered from three months after surgery. The olfactory function was evaluated by the Korean Version of the Smell Test (KVSS), which showed an improvement from a preoperative value of 9 to a postoperative value of 13 at three months. Endoscopy and computed tomography showed no intranasal cerebrospinal fluid leakage (Table 1).

Discussion

Ossifying fibroma is well-demarcated benign tumors with an aggressive tendency to locally destroy surrounding tissue. It most commonly occurs in the mandible. Other sites of origination for ossifying fibroma are associated with a more aggressive and high recurrence rate [1]. Radiologically, it appears as a rounded mass with a radiolucent center and an eggshell-like rim [2,3]. And histologically it remains with an osteoblastic rim, and can be observed within a fibrous stroma composed of glial and fibroblastic

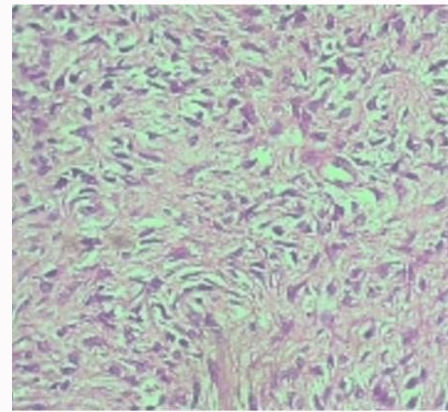


Figure 3: Histopathologic photomicrograph of the ossifying fibroma. (20x magnification with hematoxylin-eosin stain).

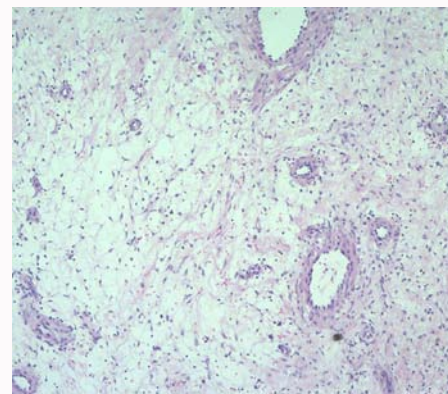


Figure 4: Histopathologic photomicrograph of the hemangioma. (40x magnification with hematoxylin-eosin stain).

cells [4]. Ossifying fibroma is classified into two types, conventional ossifying fibroma and juvenile ossifying fibroma [5]. Conventional ossifying fibroma mainly occurs in adults [1], juvenile ossifying fibroma presents in children or adolescents and tends to be more aggressive, destructing adjacent structures such as skull base, orbit bony wall, and paranasal cavity [5]. Nasal hemangioma was first reported as human botryomycosis by Poncet and Dor [6,7]. In the papers, it occurred a female predominance and was presented in the 30 to 40 years old ages [6,8]. Nasal hemangioma is usually showed enhancing soft tissue density mass like lesion with associated fluid in the radiological computed tomography. Also, the histopathological finding of hemangioma presented hemorrhagic fibro-collagenous tissue lined by squamous epithelial lining and a collection of thinned wall vascular channels in which some are dilated lined by flat and bland epithelium (Figure 4) [6,9].

In view of the similar clinical features of headache and unilateral nasal obstruction, differential diagnosis should include other neoplasms, such as fibrous dysplasia, cementoblastoma. Fibrous dysplasia involves the change of normal bone with fibrous tissue including immature woven bones. The adjacent framework of fibrous dysplasia bony lesion is maintained, as fibrous dysplasia grows slowly [2]. In contrast, ossifying fibroma presents concentric tumor-like growth [2]. Another differential diagnosis is cementoblastoma, which is a cementum-forming benign neoplasm characterized by constant proliferation. Cementoblastoma is typically locates on the tooth





	POD#1W	POD#1M	POD#2M	POD#6M
OPD based- Endoscopic Finding				
C/C relief score [VAS]	8	6	2	0
Olfactory Function Evaluation [KVSS]	NA	9	13(cf. POD#3M)	15

Table 1: Outpatient department follow-ups after ossifying fibroma removal.
 POD: Postoperative Department; W: Week; M: Month; C/C: Chief Complaint; OPD: outpatient Department; VAS: Visual Analogue Scale; KVSS: Korean Version of Sniffin' Sticks test; cf.: confer; NA: Not applicable

root and represents cementocytes and basophil bone depositions in mature bone trabecula without dentin or enamel tissue [2]. Surgical complete resection is the gold standard treatment of choice. The transnasal endoscopic approach, which preserves the integrity of craniofacial bones and structures, is the mainstay of ossifying fibroma treatment. Various surgical options are available, including transcranial, transorbital, and transfacial methods, with open resection considered in cases of extensive disease. Open resection is usually followed by facial and orbital reconstruction. Intraoperative bleeding is significantly reduced when a bony shell is present adjacent to the skull base. Staged surgery may be considered for addressing residual lesions during surveillance.

Conclusion

In conclusion, this case highlights the importance of taking into account the preoperative meticulous evaluation for risk factors such as massive bleeding and the mass lesion adjacent to skull base. To treat ossifying fibroma, the differentiation related to ossifying fibroma is critical. The standard treatment of ossifying fibroma is surgical approach. During the surgical treatment, adequate hemostasis and navigation imaging should be considered to do complete tumor removal and control the remnant tumor. And, postoperative-monitoring is essential for tumor recurrence.

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