Case Report

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Juvenile psammomatoid ossifying fibroma with skull base and orbital wall invasion: a rare case report*

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Abstract

Juvenile ossifying fibroma is an uncommon, benign, fibro-osseous neoplasm with aggressive local growth. This tumor is distinguished from other fibro-osseous lesions especially by its age of onset, aggressive behavior, and clinical presentation. Two microscopic types have been described: trabecular and psammomatoid variants. In this case report, we presented a young male patient with juvenile psammomatoid ossifying fibroma involving the sinonasal region, skull base and the orbit, which was treated with endoscopic surgery.

Keywords: Endoscopic sinus surgery, juvenile ossifying fibroma, psammomatoid variant.

Özet: Kafa tabanı ve orbita duvarına invazyon gösteren jüvenil psammomatoid ossifiye fibrom: Ender görülen bir olgu sunumu

Jüvenil ossifiye fibrom nadir görülen, benign ancak lokal agresif büyüme paterni gösteren bir fibroosseöz lezyondur. Diğer fibroosseöz tümörlerden, özellikle hastalığın başlangıç yaşı, agresif davranışı ve klinik presentasyonu ile ayrılır. İki mikroskopik tipi tanımlanmıştır; trabeküler ve psammomatoid varyant. Çalışmamızda, genç bir erkek çocukta sinonazal bölge, kafa tabanı ve orbitayı tutan ve endoskopik olarak tedavi edilen bir psammomatoid ossifiye fibrom olgusunu sunduk.

Anahtar sözcükler: Endoskopik sinüs cerrahisi, jüvenil ossifiye fibrom, psammomatoid varyant.

Ossifying fibromas are benign tumors with locally aggressive behavior and have the potential risks for bone destruction and high recurrence.[1] Juvenile ossifying fibroma (JOF) is an uncommon lesion that has been distinguished from other ossifying fibromas by age, frequent site of involvement, and clinical behavior.[2] This tumor is quite aggressive and has a high predisposition to relapse.[3] The term JOF is utilized in the literature for a description of two different histopathologic patterns of ossifying fibroma, which are trabecular juvenile ossifying fibroma (TJOF) and psammomatoid juvenile ossifying fibroma (PJOF). The term psammomatoid juvenile ossifying fibroma was first introduced by Margo et al. in 1985.[4] Other names used in the literature include juvenile aggressive psammomatoid ossifying fibroma, psammo-osteoid-fibroma, and psammous desmo-osteoblastoma. PJOF mostly originates from the sinonasal region and orbital bones at young age and shows an infiltrative growing pattern. It has a strong tendency for recurrence.[5] This clinical entity is often confused with malignancy owing to its aggressive and osteolytic nature.[6] In this study, we reported a case of PJOF involving the sinonasal region, the skull base and medial orbital wall which was treated with endoscopic surgery under the guidance of navigation.

Case Report

A 19-month-old male patient was consulted with the complaints of a swelling in the left infraorbital region, facial asymmetry, left nasal obstruction and breathing difficulties for two months. Past history revealed that he had undergone magnetic resonance imaging (MRI) due to a mild

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facial swelling in another hospital 14 months ago which has been reported as non-diagnostic.

On rhinoscopic examination, there was an expansile lesion, involving and obstructing the left nasal cavity. This lesion was firm and exophytic as well. Eye movements to lateral and superior direction were restricted on the left side with an intact light reflex. A painless swelling on the left infraorbital region had caused a facial asymmetry. However, the general health condition of the patient was good. No weight loss, fever or night sweating was reported.

MRI findings showed a 48×36 mm lesion which had obliterated the left nasal cavity, left maxillary, ethmoid and sphenoid sinuses, destroyed the skull base, and invaded the medial and inferior rectus muscles and the orbit inferomedially. It had also pushed the globe (Fig. 1). An incisional biopsy was performed for histopathological diagnosis and revealed spherical or curved ossicles resembling psammoma bodies dispersed in a cellular fibroblastic stroma. Morphological findings were consistent with juvenile ossifying fibroma (Fig. 2).

Functional endoscopic sinus surgery under the guidance of navigation was planned. The left nasal cavity, left maxillary antrum and posterior wall of the antrum, medial wall of the left orbit, left sphenoid sinus, the septum of the sphenoid sinus and the skull base were observed to be invaded by the tumor. The dura was intact. The lesion was removed endoscopically piece by piece from all compartments. Macroscopic inspection revealed multiple gray-white fragments. Post-operative recovery was uneventful and the patient was discharged one week after surgery.

Short-term (one month after the surgery) MRI findings revealed that the tumor was substantially removed. Local hypointense areas in left frontoethmoidal recess and maxillary sinus were regarded as the fragments of Nasopore (Polyganics, Groningen, Netherlands) which is an absorbable dressing material used during the surgery for hemostasis (Fig. 3).

Discussion

Juvenile ossifying fibroma is a rare fibro-osseous lesion of the craniofacial skeleton described by the onset before 15 years of age, the tumor location, radiological presence, and high-level recurrence potential. This benign tumor may present as one of two histological patterns: PJOF and TJOF.[7] Some authors previously defined the adult form as cemento-ossifying fibroma and the juvenile form as ossifying fibroma.[8] However, this nomenclature has been aborted, and now denominated as "ossifying fibroma"

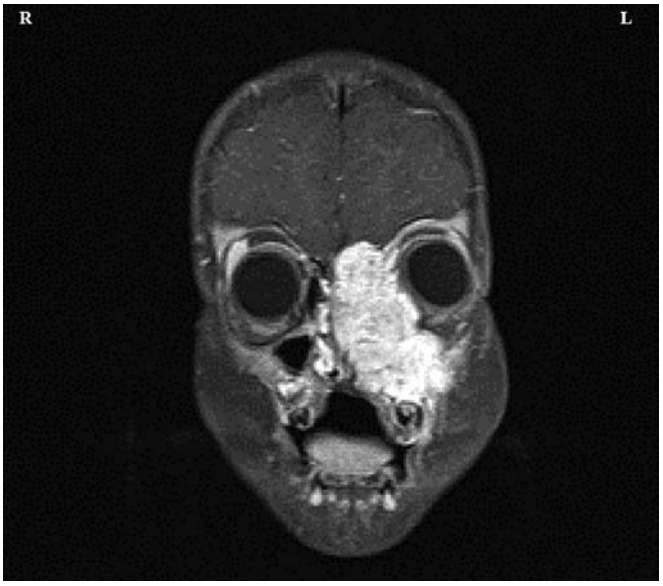

Fig. 1. Pre-operative MRI image of the tumor located in the left sinonasal region. The left anterior skull base, medial orbital wall, maxillary and frontoethmoidal sinuses, and left dento-alveolar arc are invaded by a well-demarcated, heterogeneous, expansile lesion.

owing to the comparable histological features.[9,10] Three types of ossifying fibromas have now been described: (1) classical ossifying fibroma, (2) PJOF, and (3) TJOF.[11] The psammomatoid type is reported as more aggressive than a trabecular variant, and has a high tendency to recur.[12]

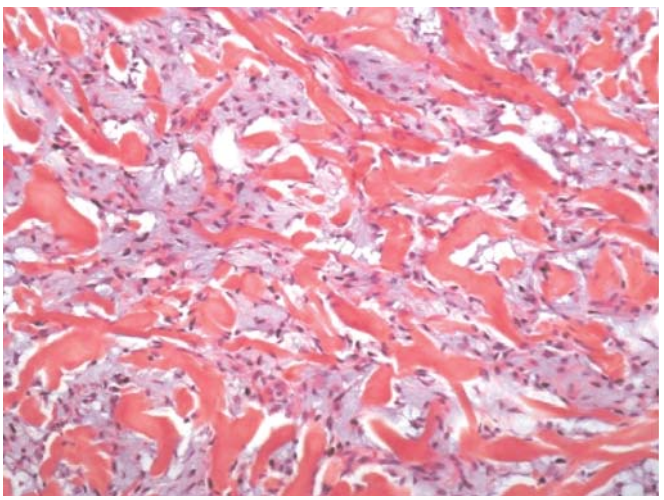

Fig. 2. The histopathological image of JPOF demonstrates small ossicles resembling psammoma bodies in fibroblastic stroma (hematoxylin and eosin stain with x200 magnification). [Color figure can be viewed in the online issue, which is available at www.entupdates.org]

The JOF, as the name indicates, has a high incidence in children and young adults. It was reported to be seen predominantly in males between first and second decades of life.[4] The presented case was also male and 19-month-old.

The etiology is unknown. However, the primitive mesenchymal tissue, which is located relatively more in mandible, maxilla, ethmoid sinuses and skull base than the other locations, was thought to be the origin of this rare tumor. On the other hand, the overproduction of the myxofibrous cellular stroma forming the septa of paranasal sinuses was considered to be responsible.[13]

Psammomatoid variant mostly affects the sinonasal and orbital bones of the skull, whereas TJOF predominantly occurs in mandible and maxilla.[12] In our case, sinonasal region, skull base, and orbital medial wall were involved.

JOF causes asymptomatic facial swelling. Even if the growth duration depends on the site and aggressiveness of the lesion, these tumors do not demonstrate a long-standing evolution as other fibro-osseous lesions.[3]

The presence of cellular fibrous stroma, immature bony strands, and cement particles is the characteristic histological appearance of JOF.[8] The juvenile psammomatoid variant is characterized by numerous small, round ossicles, or "psammomatoid" bodies embedded in a cellular fibrous stroma.[14]

Radiographically, the lesion can be radiolucent, mixed or radiopaque, depending on the degree of calcification. Although JPOF is not a capsulated tumor, it can be separated from the surrounding bony structures by radiopaque borders. This radiological feature helps to differentiate it from fibrous dysplasia.[15] The present case demonstrated a mixed radiopaque and lucent lesion.

Fibrous dysplasia should be the first to be considered in the differential diagnosis. The rapid growth process and well-delineated radiographic borders differentiate JOF from fibrous dysplasia.[16] The diseases including aneurysmal bone cyst, osteosarcoma, osteoblastoma and cementoosseous dysplasia should be thought in the differential diagnosis as well.

Surgical resection is suggested for the treatment of JPOF owing to its aggressive nature and high recurrence rate which was reported about 30–60%.[17] Endoscopic sinus surgery, lateral rhinotomy, craniotomy or mixed (endoscopic and external) approach can be chosen for the resection of this tumor varying from case to the case. In the presented patient, endoscopic removal was preferred. Due to the age of the case, no aggressive surgery, but close follow-up was decided. Recurrence may occur by reason of difficulty in complete resection caused by the location and the infiltrative borders of the tumor. Long-term follow-up is needed owing to its aggressive nature and high recurrence rates. Radiotherapy is contraindicated because of the risk of malignant transformation and unfavorable effects in children.[15]

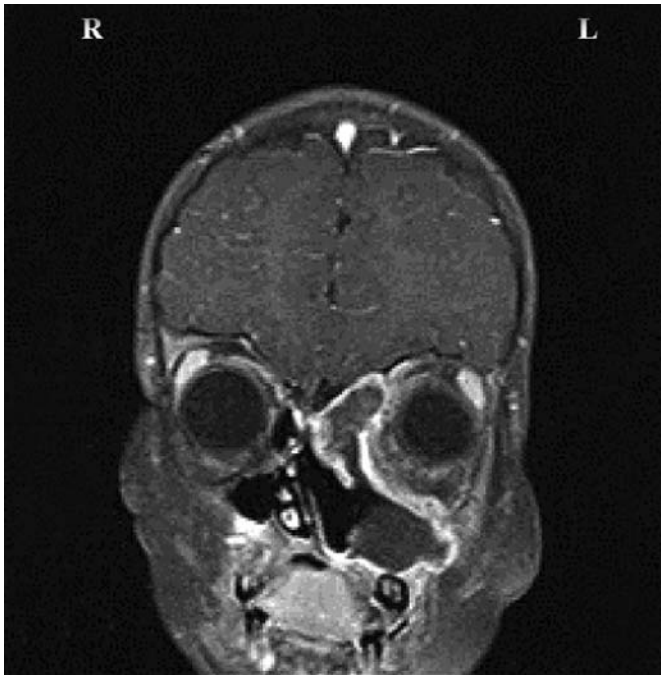

Fig. 3. Post-operative MRI image of the patient one month after the endoscopic sinus surgery. The tumor was substantially removed. Absorbable dressing material fragments used for hemostasis during surgery can be seen as hypointense areas in maxillary sinus and frontoethmoidal recess.

In conclusion, JOF is a rare, rapidly growing and local aggressive benign lesion which is often confused with malignancy. Surgical excision is suggested for the treatment. Long-term follow-up is necessary due to the high recurrence rate of this clinical entity.

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Conflict of Interest: No conflicts declared.

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