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CASE REPORT

Juvenile psammomatoid cemeto-ossifying fibroma: Two cases

Ja Hee Seo,[1] Doo Hee Han,[2] Chae-Seo Rhee[2] and Sung-Hye Park[1]

[1]Department of Pathology, Seoul National University Hospital and [2]Department of Otorhinolaryngology, Head and Neck Surgery, Seoul National University College of Medicine, Seoul, Korea

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Correspondence
Sung-Hye Park, MD, PhD, Department of Pathology, Seoul National University College of Medicine, 28 Yeongeon-dong, Jongno-gu, Seoul 110-799, Korea. Email: shparknp@snu.ac.kr

ABSTRACT

Juvenile ossifying fibroma is a variant of ossifying fibroma, commonly involving sinunasal tract of children. It is an aggressive condition that leads to the destruction of adjacent structures. Distinction from other fibro-osseous lesions is important. Juvenile ossifying fibroma has two histologic subtypes that are psammomatous and trabecular type. We present herein two cases of juvenile psammomatoid ossifying fibroma occuring in a 22 month old and a 5 year old child, which arose in the bilateral maxillary sinuses and led to bulging of the cheek and nasal obstruction. The tumor in case 1 could not be completely excised due to its huge size and massive destruction of the bilateral maxilla. The tumor in case 2 was completely excised through staged operations. Tumors in both cases were diagnosed as juvenile psammomatoid cement-ossifying fibroma by the histology. Here, we report two cases from Korea and describe psammomatoid morphology and how to differentiate it from other fibro-osseous lesions.

INTRODUCTION

Ossifying fibroma (OF) is a rare and unique fibro-osseous neoplasm described by Bendet *et al.*,[1] that commonly develops in the craniofacial bone, especially in sinonasal tract and orbital area. The 1972 World Health Organization (WHO) classification separated cementifying fibroma from ossifying fibroma, however, in the 1992 WHO classification, it was considered to be a histologic variant of ossifying fibroma.[1] Clinically, the juvenile form of cement-ossifying fibroma (COF) is a subgroup of COF mainly occurring in childhood and adolescence (ages 5–15 years).[2] Recurrence is more common in juvenile COF (JCOF) than in other COFs.[1] OF may be indolent or aggressive. The JCOF subgroup behaves aggressively resulting in destruction of the adjacent structures and sometimes leads to facial asymmetry.[3]

COF is described variously as OF, cementifying fibroma, psammomatoid OF, trabecular OF, juvenile (psammomatoid) OF, etc., which produces confusion. However, if we understand each name as being linked to the histological features, it is simple to distinguish various names and subtypes. The psammomatous and trabecular subtypes of COF have been well documented. The psmmomatous subtype has spherical or ovoid bony component, which resembles psmmoma bodies. Osteoblastic rimming may or may not be present. Some ossicles are calcified, having basophilic center and eosinophilic fringe with lamellation. The trabecular subtype possesses irregularly shaped trabeculae of immature bone with or without osteoblastic rimming instead of spherical or ovoid bone. Sometimes these trabeculae are cellular and show anastomosing pattern. The fibrous components of the JPOF, OF, and fibrous dysplasia (FD) have similar features.[4] We report herein two rare cases of JPOF that involved the maxilla in young children.

CASE REPORTS

Case 1

A 22-month-old girl was referred to the Department of Otorhinolaryngology of Seoul National University Hospital and presented with a 3-month history of persistent nasal obstruction, rhinorrhea, snoring and sleep apnea. Physical examination revealed bulging of bilateral cheeks and hard palate. In preoperative computed tomography (CT) scans, two ossifying lesions were observed fully filling up the alveolar process and expanding to the nasal cavity, resulting in the nasal obstruction (Fig. 1). CT showed symmetric expansile lesions with ground glass opacity. The lesions involved the root of a maxilla causing the displacement of an unerupted tooth.

Endoscopic biopsy was performed. Intraoperatively, lesions were noted bilaterally bulging into the inferior meatus (Fig. 2). The lesion in the right nasal cavity was incised after elevating the inferior turbinate. The inner component of the mass was whitish and cartilaginous. The diagnosis of juvenile psammomatoid cement-ossifying fibroma (JPCOF) was confirmed (Fig. 3a).

Histologically, the tumor was composed of numerous psammoma-like small ossicles with a thick collagenous rim in the fibrous stoma.

Case 2

A 5-year-old boy presented with a 3-month history of swelling in right cheek. CT showed a mass at the right maxillary sinus that had destroyed the inferior orbital wall and alveolar process (Fig. 4a). The mass had calcifications and small cystic changes within the inner portion. Biopsy was performed and osteoblastoma diagnosed. Histological examination of the excised tumor revealed JCOF (Fig. 3b).

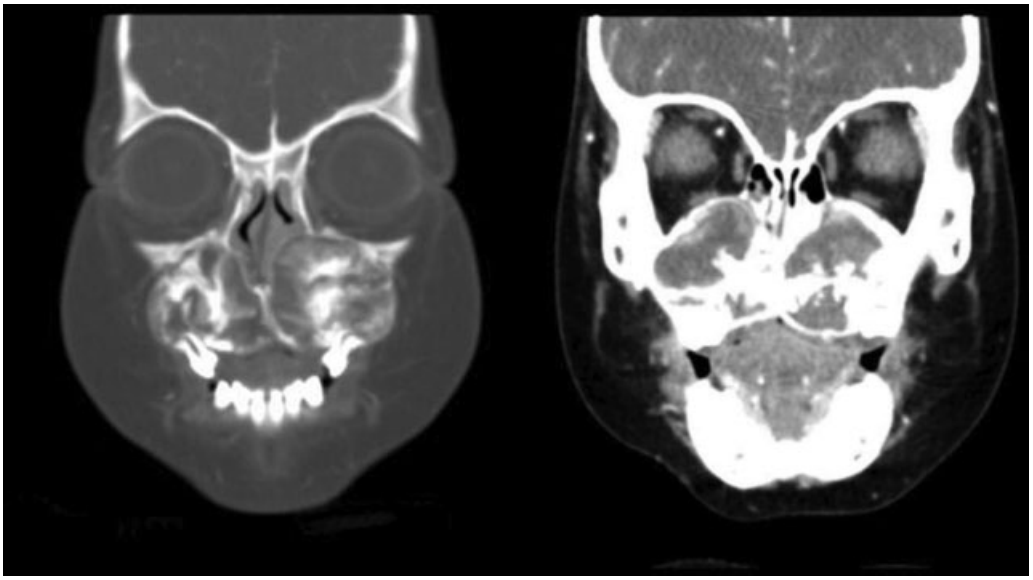

Figure 1 Preoperative coronal computed tomography scans showing two expansile mass lesions involving nearly entire maxilla bilaterally.

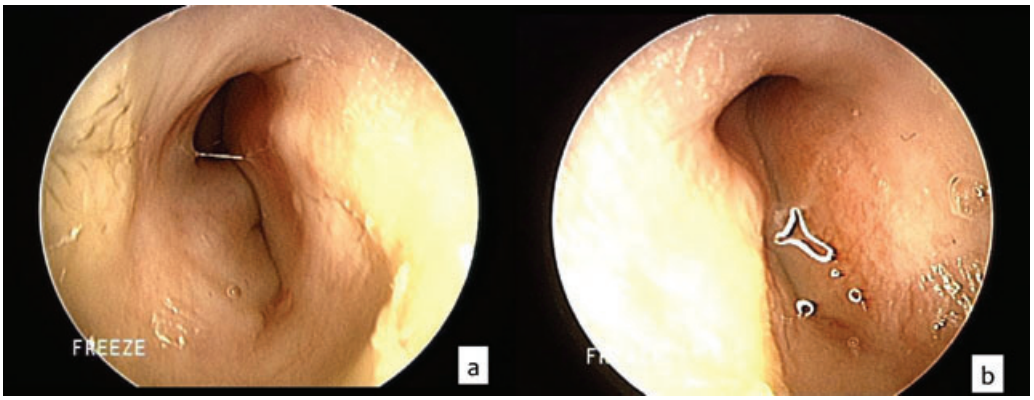

Figure 2 Endoscopic examination showing bulging mass lesions in both nasal cavity. (a) Right nasal cavity and (b) left nasal cavity.

Excision of the mass was contemplated using the Caldwell–Luc approach, but complete removal was not possible due to massive hemorrhage. Six months later, the Caldwell–Luc approach was successfully carried out after embolization.

Microscopic examination of the masses showed psammomatoid osteoid and basophilic cementicles without definite osteoblastic rimming. These psammomatoid ossicles were surrounded by fibroblast-like spindle cells and osteoclast type giant cells (Fig. 3).

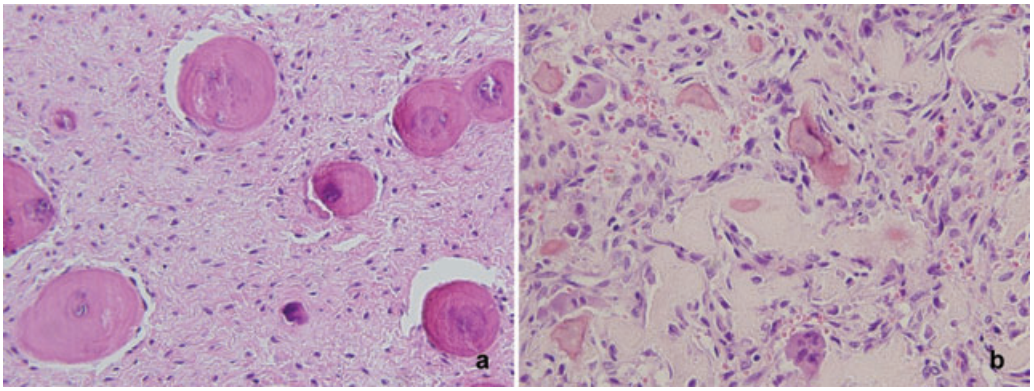

Figure 3 Microscopic findings of tumor. (a) Psammomatoid ossicles (b) and osteoclastic giant cells surrounded by fibroblast-like spindle cells were observed.

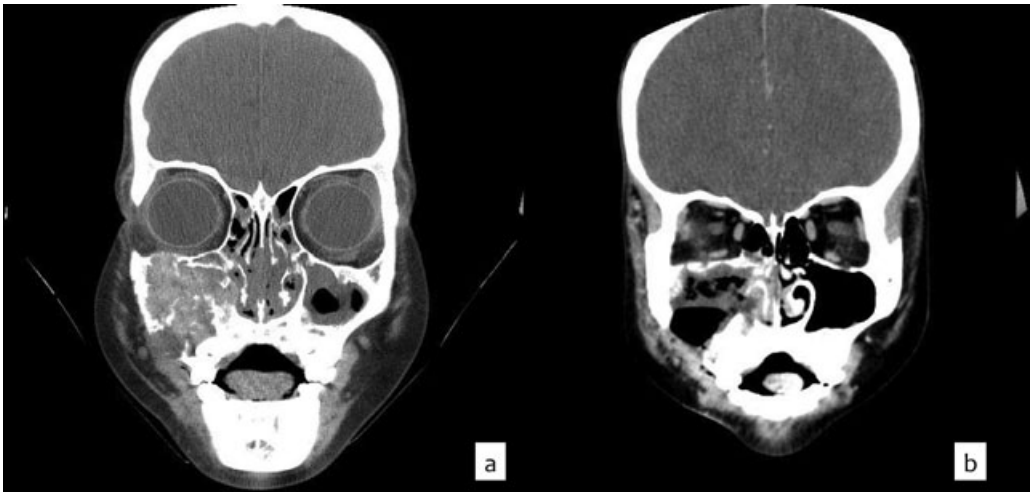

Figure 4 Computed tomography (CT) scans before and after surgery. (a) Preoperative coronal CT scans showing bone tumor with the destruction of adjacent structures including the inferior orbital wall and alveolar process. (b) CT scans a week after surgical excision showing no residual mass with fluid collection.

Few inflammatory cells were noted to be infiltrating spindle cells.

Immunohistochemically, the spindle cells in the non-osseous component were non-reactive to smooth muscle actin and epithelial membrane antigen. Immunistaining of CD56 to detect osteoblast showed non-specific staining pattern. CD68, the marker for osteoclast and macrophage stained the perivascular area. The spindle cell component was more cellular and ossification was more prominent in the tumor in case 2 than that in case 1.

DISCUSSION

Benign fibro-osseus lesions of the maxilla and mandible are classified into three categories: fibrous dysplasia, dysplastic lesion and fibro-osseous neoplasm. In the WHO classification, cemetifying fibroma and OF are considered to be a spectrum and are therefore referred to as ossifying fibroma.[1] Cemetum is a specialized calcified substance covering the root of teeth. There are three type of cementum; acellular, cellular and afibrillar. The acellular cementum lacks cellular components. The chemical makeup of cementum is similar to that of bone, but is not vascularized. Often the osseous component, which may be mature or immature, appears bluish and acellular, similar to a cementum, hence the name OF is coined.

COF is a benign, monostotic well-circumsribed unilocular or multilocular fibrous-osseous tumour, known to arise from the periodontal ligament. It is more common in 30–40-year-old women, with age range of 10–59 years.[5,6] The circumscription of the tumor differentiates it from fibrous dysplasia because FD lesions are ill-defined.[4] COFs arise most frequently in the mandible (about 70% cases) but a significant percentage of cases (22%) are found in the molar region of the maxilla, ethmoidal and orbital regions and exceptionally in petrous bone.[7,8] Radiologically, the COFs are radiolucent in 53% of cases, a sclerotic radio dense in 7% and mixed or mottled appearance in 40% of the cases.[8]

COF is subclassified based on age; that is, JCOF and conventional. These entities possess two types of typical morphologic features: psammomatoid (ovoid ossicles) or trabecular bone. Psammomatoid COF has ovoid ossicles while the trabecular COF has cellular or acellular trabecular bones.[9]

JCOF can be distinguished from adult type by its clinical, radiological, and pathological characteristics. JCOF mostly involves the maxilla and paranasal sinus of children or adolescence and locally aggressive.[1] It does not present without clinical symptoms, such as pain. Because of its aggressiveness, it is often mistaken for osteosarcoma, osteoblastoma, or other malignant bone tumors.[4] Thus, it is crucial to distinguish it from other malignant bone tumors or fibro-osseous lesions.

The etiology of JCOF has not been fully understood. Genetic differences in fibro-osseous lesion of craniofacial bone are not well established, but there are reports that COFs were related to non-random break points at Xq26 and 2q33.[10] This may be used to differentiate COFs from figrous dysplasia, which is associated with mutation of the alpha-subunit of the stimulatory G protein gene.[11] The treatment of choice for COF is complete surgical resection[12] and is usually cured by this treatment. Incomplete resection of the tumor may result in recurrence.

Our two cases of JPCOF occurred in young individuals (22 months, 4 years) and presented with the expansile masses, involving maxilla, presented with asymmetric swelling of cheek or nasal symptoms. In the first case, complete excision via staged operation (excision of tumor at right side first and followed by that at left) was contemplated, while case 2 underwent excision via staged operation after embolization.

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