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Review Article

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Aneurysmal bone cyst of the maxillary sinus secondary to fibrous dysplasia: Report of a rare case and review of the literature

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Abstract

ABC developing within FD in head and neck region is quite rare. Clinical presentation of FD and ABC depends on the location and scope of content. The majority of reported cases were seen in male children and adolescents. Complete excision of these lesions is the recommended treatment.This report is about a 27 year-old male patient who presented with a mass in hard palate treated with total excision.

Introduction

Fibrous Dysplasia (FD) is a benign skeletal disease in which medullary bone is replaced with fibrocellular tissue. It results from abnormal fibroblast development [1]. Aneurysmal Bone Cysts (ABC) are relatively rare, benign vascular lesions and also may exist secondary to the pathological bone lesions [2].

FD in combination ABC that appears as a symptomatic or asymptomatic mass is quite rare. Clinical presentation of FD and ABC depends on the location and scope of content. The majority of reported cases are seen in male children and adolescents. Complete excision of these lesions is recommended.

Case report

27 year old male patient admitted to our clinic with complaints of swelling in hard palate and toothache. Medical history was unremarkable, and general health condition was well. On physical examination, a 4x5 cm soft mass was seen on the middle part of the hard palate. The overlying mucosa was intact and bony consistency of the hard palate could not be palpated. Nasal endoscopy was unremarkable.

Paranasal sinus CT scan revealedthat the lesion was originating from the inferior wall of the maxillary sinus and was surrounding the left posterior molar teeth. The central part of the lesion had soft tissue dansity, periferal areas had ground glass opacity and the nature of the lesion was expansile (Figure 1). Age and radiological findings of the patient were compatible with the fibrous dysplasia.

Caldwell-Luc antrostomy was performed and the lesion that filled the maxillary sinus and expanding to hard palate was totally excised with blunt dissection. Inferior antrostomy was done and stabilisated with a rubber drain. Histopathological examination was reported as aneurysmal bone cystsecondary to fibrous dysplasia with woven bone formation and short spindle cells within the stroma (Figures 2 and 3).

Literature review and discussion

Fibrous dysplasia which was first described by Lichtenstein [3] constitutes 2,5 % of all bone tumors and %7 of benign bone tumors [4]. It is often seen in young male patients. FD can present as monostotic (%70) or polystotic (%30) disease and may be a component of McCune-Albright syndrome [5]. At the craniofacial area; maxilla, mandible, frontal and temporal bones are the most frequent sites of ivolvement. CT images usually reveal extensive diploic spaces, enlargement and ground glass opacity of the affected bones and MRI shows typically low signal intensity on T1 and T2-weighted images [6].

ABC can be seen about 1.4% of all bone tumors but only 3-6% of them present in the cranium [7]. ABC mostly affect patients under under 20 years without gender difference [8]. Fluid-filled cavities are common and can be detected on CT or MR images [9]. On histopathologic examination; ABC present with blood-filled cavernous spaces which are usually seperated by fibrous tissue.

This report presents a case who was diagnosed as concominant FD and ABC. The occurance of ABC in combination with FD in the head and neck region is extremely rare. Up to date only several cases has been reported and most of these cases the disease is located on the skull base with only 3 cases of maxilla involvement. The cases of concomitant FD and ABC on the sinonasal region are summarized in Table 1 [10-14].

The presentation of these lesions depends on the sites of involvement, growth rate and anatomic structures of the involved region. Craniofacial lesions usually present with symptoms resulting

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Table 1. Fibrous dysplasia with aneurysmal bone cysts at the sinonasal region

Reference	Age (years) Gender	Symptoms	Localization	Radiological findings	Treatment	Following period
Saito *et al.* 1998 [10]	11/M	Nasal obstruction for 1 year	Nasal cavity and sphenoid bone	CT and MRI, irregular multilobulated tumor	Surgical excision	No recurrence for 3.5 years follow-up
Skaladzien *et al.* 2008 [11]	16/M	Rhinosinusitis and epistaxis	Right maxillary sinus	Large cystic lesion	Surgical excision	No recurrence for 9 months follow up
Pasquini *et al.* 2002 [12]	5/M	Chronic rhinosinusitis for 2 years	Right maxillary sinus	CT: Cystic lesion	Transnasal endoscopic surgery	No follow-up
Lin *et al.* 2004 [13]	18/M	Mass with headache	Left frontal bone	CT: Cystic spaces	Surgical excision	No follow- up
Terkawi *et al.* 2011 [14]	7/F	Left nasal obstruction and left eye blindness	Sphenoid and ethmoid bones	CT-MR: Cystic lesion	Endonasal - cranial resection	Recurrence was revealed after 5 months
Our case	27/M	Toothache and mass at hard palate	Left maxillary sinus and bone	CT: Expansile mass in the left maxillary sinus	Surgical resection	No recurrence for 2 years follow-up

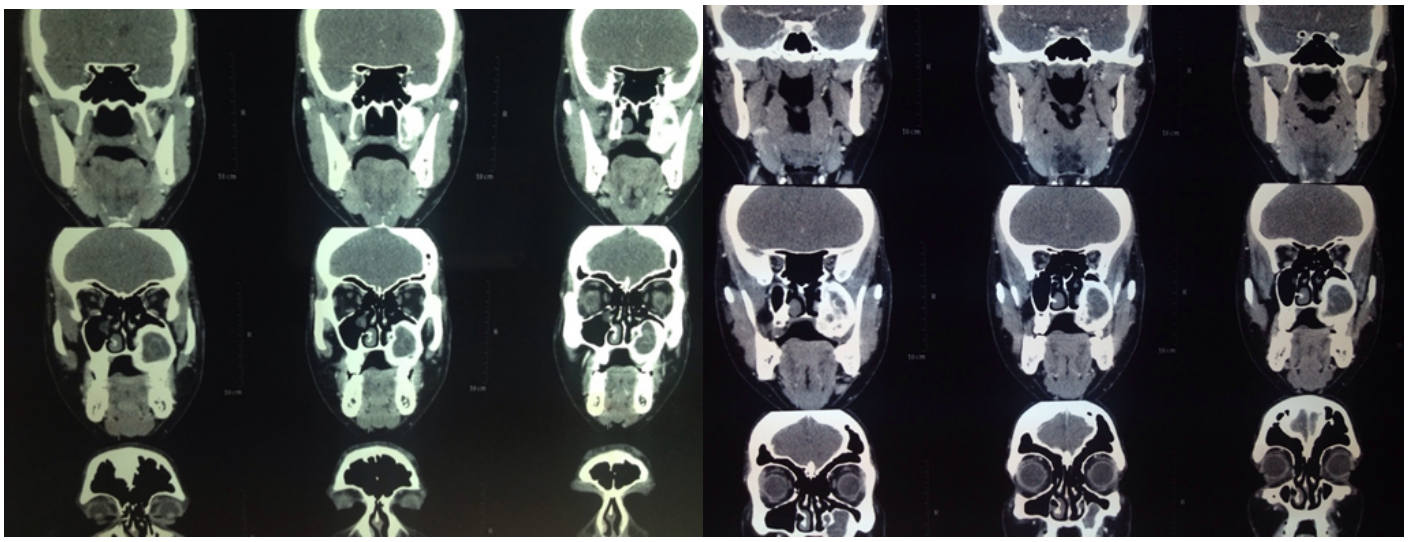

Figure 1. Expansile lesion was originating from the inferior wall of the maxillary sinus. Central soft tissue dansity and peripheral ground glass opacity can be seen.

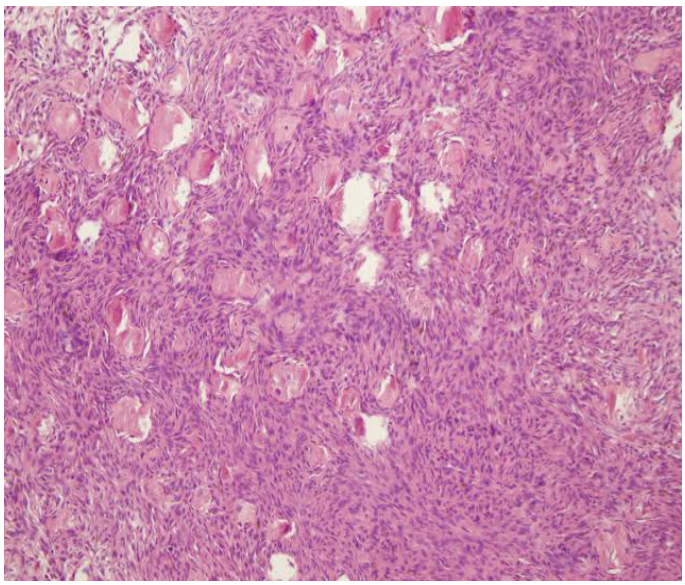

Figure 2. Fibrous dysplasia shows characteristic pattern of woven bone formation and short spindle cells within the stroma (H&EX200).

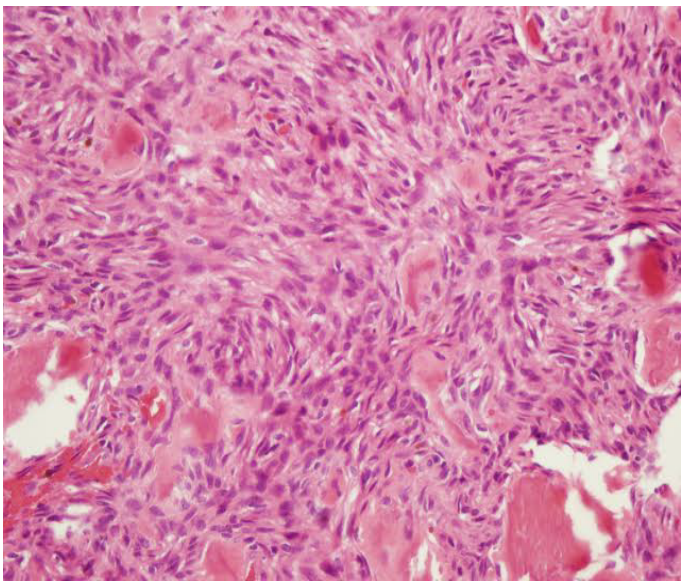

Figure 3. Bland oval to spindle-shaped stromal cells without cytologic atypia surrounds woven bone (H&EX400).

from the mass effect of the lesion. The lesions may present with painless mass or may cause symptoms specific to site such as nasal obstruction, headache (Saito et. al.'s patient; 10) and loss of vision (Terkavi et. al.'s patient); 14). Our case presented with mass over the hard palate. Consistent with other reports the growth pattern of our case was slow without any compression over vital structures.

Diagnosis of FD combined with ABC depends on radiologic and pathologic findings. On radiology the typical appearance of FD

as ground glass opacity at the periphery of the lesion was detected on CT images combined with cystic soft tissue dansiy in the center. Intraoperative abundant bleeding rouse the suspect of ABC and postoperative definitive pathology confirmed the diagnosis ABC associated with FD. We believe that typical findings of FD combined with cystic spaces on CT images may be helpful in preoperative differential diagnosis and precautions about bleeding during surgery may be undertaken accordingly.

Optimal treatment of these lesions is total excision. Other treatment modalities are arterial embolization, sclerotherapy, cryotherapy, radiotherapy or combination of these methods but recent studies showed that radiotherapy alone has the risk of post-irradiation sarcoma [15]. In secondary ABC's, such as our case, the treatment plan should be set according to the site of primary lesion. Most of the reported cases (concomitant FD with ABC) treated with surgical excision and the results are quite good. We treated our case by total surgical excision. The lesions impingement symptoms and findings or patients with cosmetic problems should be treated by surgery and as fibrous dysplasia has a potential of malignant transformation (0.5% of patients with monostotic FD; 4% of patients with polystotic FD can develop malignant transformation) [16] the patients should be closely monitored. In our case after two years follow-up the patient is well without reccurence.

In conclusion although combination of FD and ABC is rare, awareness of such cases may lead preoperative suspect and preoperative and intraoperative precautions on bleeding from ABC.

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