Case Report

A Rare Presentation of Ossifying Fibroma with a Secondary Aneurysmal Bone Cyst in the Maxillary Sinus – A Case Report

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ABSTRACT

This case report describes a rare occurrence of coexisting ossifying fibroma (OF) and aneurysmal bone cysts (ABC) in the maxillary sinus (MS) of a 22-year female patient. Patient reported to Dental Clinics of College of Dentistry, Qassim University in December 2022. The initial presentation included proptosis and diplopia in the left eye, which prompted further investigations. Imaging revealed a large complex solid and cystic tumor mass extending into the orbit from the left MS. Endoscopic incisional biopsy confirmed the diagnosis of a cyst of the MS, characterized by a giant cell-rich benign fibro-osseous lesion suggestive of OF, with a secondary ABC. This case highlights the importance of considering these rare entities and their coexistence in the differential diagnosis of MS lesions, emphasizing the role of a biopsy in confirming the diagnosis and guiding appropriate management strategies.

Key Words: Ossifying Fibroma, aneurysmal bone cyst, diplopia, Proptosis

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INTRODUCTION

Ossifying fibroma (OF) is a rare and non-cancerous tumor which have seen primarily affects the jaw and facial skeleton.[1] It involves the abnormal growth of fibro-osseous tissue instead of normal bone, and it can either develop slowly or exhibit local aggressiveness. The first recorded case of OF dates back to 1872 and similar cases with clinical and histopathological similarities were reported as early as 1942, using terms like central osteoma of the maxilla and OF. The term "ossifying fibroma" itself was coined in the year 1927, but its classification and variants continue to be a topic of controversy even today.[2] However, regardless of classification, surgical removal is the primary treatment once the diagnosis of OF is confirmed through biopsy. Recurrence after resection is rare, and there have been no reported cases of malignant transformation.[3]

Extra-gnathic OF mainly presents with symptoms related to its mass effect. Patients typically experience proptosis (protrusion of the eyeball) or nasal obstruction, but they may also have headaches, visual disturbances, a facial mass, or rhinorrhea (runny nose).[4]

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Occasionally, ossifying fibroma in the jawbone is accompanied by cystic changes, although this association has not been extensively documented. One of the reported cystic changes associated with OF is the presence of Aneurysmal Bone Cysts (ABCs), which are neoplastic osteolytic spaces filled with blood, and separated with fibrous septa. ABCs are commonly seen in long bones but have preference for the mandible when affecting the jaw bones. The occurrence of a maxillary OF together with an ABC is extremely rare. We present a new case of a 22-year-old female with a large OF associated with an ABC in the MS.

CASE REPORT

A 22-year otherwise healthy individual sought medical attention due to proptosis (bulging) in their left eye. After initial investigations, a large tumor was identified in the left maxillary sinus that extended into the base of the same-side orbit. The proptosis resulted in a superior and lateral deviation of the affected eye, but there was no significant impact on visual activity. Examination of the face, mouth, and neck did not reveal any obvious expansion or swelling, Mucosal changes, or cervical adenopathy. Ophthalmic examination indicated reactive pupils and mostly unaffected extraocular movement, although diplopia (double vision) was present during the left superolateral gaze. Mild exposure keratitis (inflammation of the cornea) was also observed upon closer examination. Intraocular pressure measurements using an air puff tonometer showed readings of 19 mmHg with respect to the right eye and 22 mmHg in the left eye. Fundoscopic examination revealed certain changes in the left optic nerve on a digital level.

Investigations

Radiographic Examination: To further investigate the proptosis, a CT scan with a contrast of the orbits was performed. The scan revealed a large, complex tumor mass in the left MS, predominantly cystic with a central fluid component measuring 24 Hounsfield units (HU). The solid component at the periphery ranged between 50 and 110 HU. The tumor displayed a tit-like projection into the area near the root of the left third molar, which might be the origin of the lesion. The superior cystic aspect of the mass was inseparable from the left inferior rectus muscle and abutted the inferior part of the eyeball, causing erosion of the orbital base without signs of an invasion. (Figure1-A&B)) Subsequently, an MRI was conducted, which revealed a hypointense signal on T1-weighted images and a hyperintense signal on T2-weighted images for the cystic component. The solid component exhibited heterogeneous enhancement, with the peripheral enhancement of the cystic portion. The overall dimensions of the lesion were measured as 37mm x 34mm x 34mm. (Figure 2).

Differential Diagnosis: The differential diagnosis was considered as fibrous dysplasia, ameloblastic fibro-odontoma, giant cell lesion, and OF

Incisional Biopsy: The patient's incisional biopsy, performed endoscopically by the ear, nose, and throat (ENT) specialist, led to the resolution of proptosis in the left eye and diplopia (double vision) without any subsequent development of enophthalmos. The final diagnosis revealed a cyst of the maxillary sinus, characterized by a giant cell-rich benign fibro-osseous lesion suggestive of OF, accompanied by a secondary ABC. Additionally, focal fibrosis was observed in the left maxillary wall, and another giant cell-rich benign fibro-osseous lesion suggestive of OF with a secondary ABC was identified in the inferior MS. These findings confirm the presence of these specific lesions in the MS, highlighting the coexistence of OF and ABC in the patient's case.

Histopathology: The patient underwent multiple biopsies and endoscopic excision of the cystic component. The fluid within the aneurysmal bone cyst was found to be viscous and green in color, lacking mucinous or bloody characteristics. Following the cyst excision, the proptosis and diplopia resolved. A post-excision CT scan revealed a reduction in the size of the mass, with almost complete resolution of the extension into the infraorbital region and alleviation of mass effect on the inferior rectus muscle. A histopathological examination of the biopsy confirmed the diagnosis.

Management: Surgical excision with peripheral ostectomy (Weber Ferguson approach) was performed.

Surgical Histopathology

Outcome and Follow-up: The surgical site showed no complications, remaining intact, clean, and dry. There were no signs of neurosensory deficits, indicating the procedure did not result in any nerve damage. The ophthalmic examination demonstrated improved proptosis in the left eye, with measurements showing a decrease in protrusion, further affirming the positive outcome of the treatment. A follow-up appointment was given after 1 year.

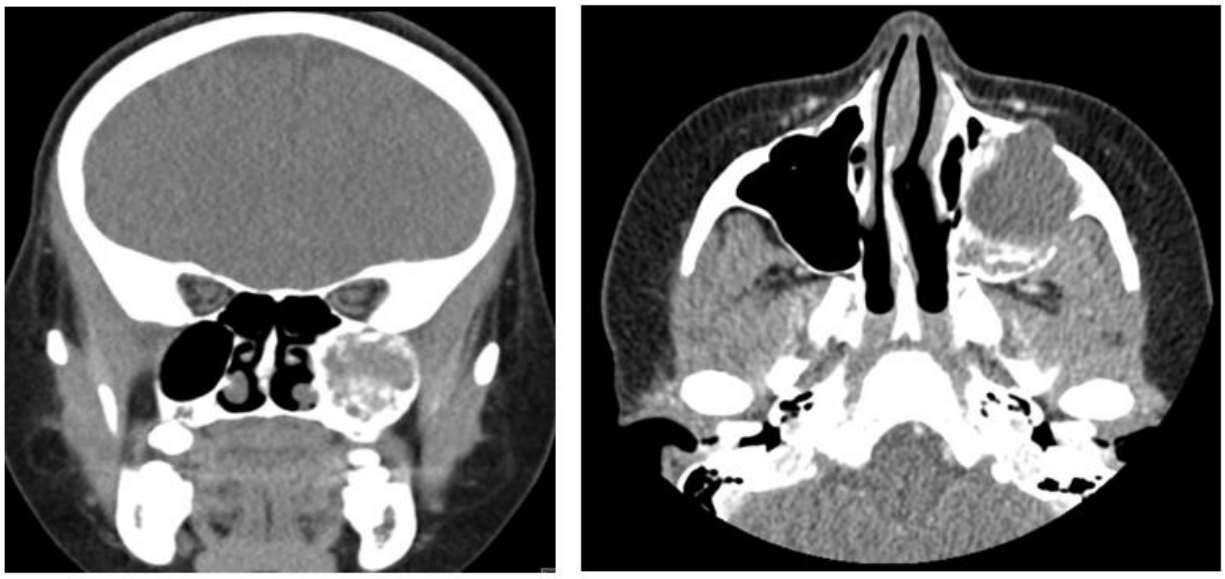

Figure No.1: CT scan of the head and neck post IV contrast showing complex solid and cystic tumor within the left maxillary sinus, extending into the ipsilateral orbital cavity with resultant ocular proptosis. (A) Coronal View (B) Axial View

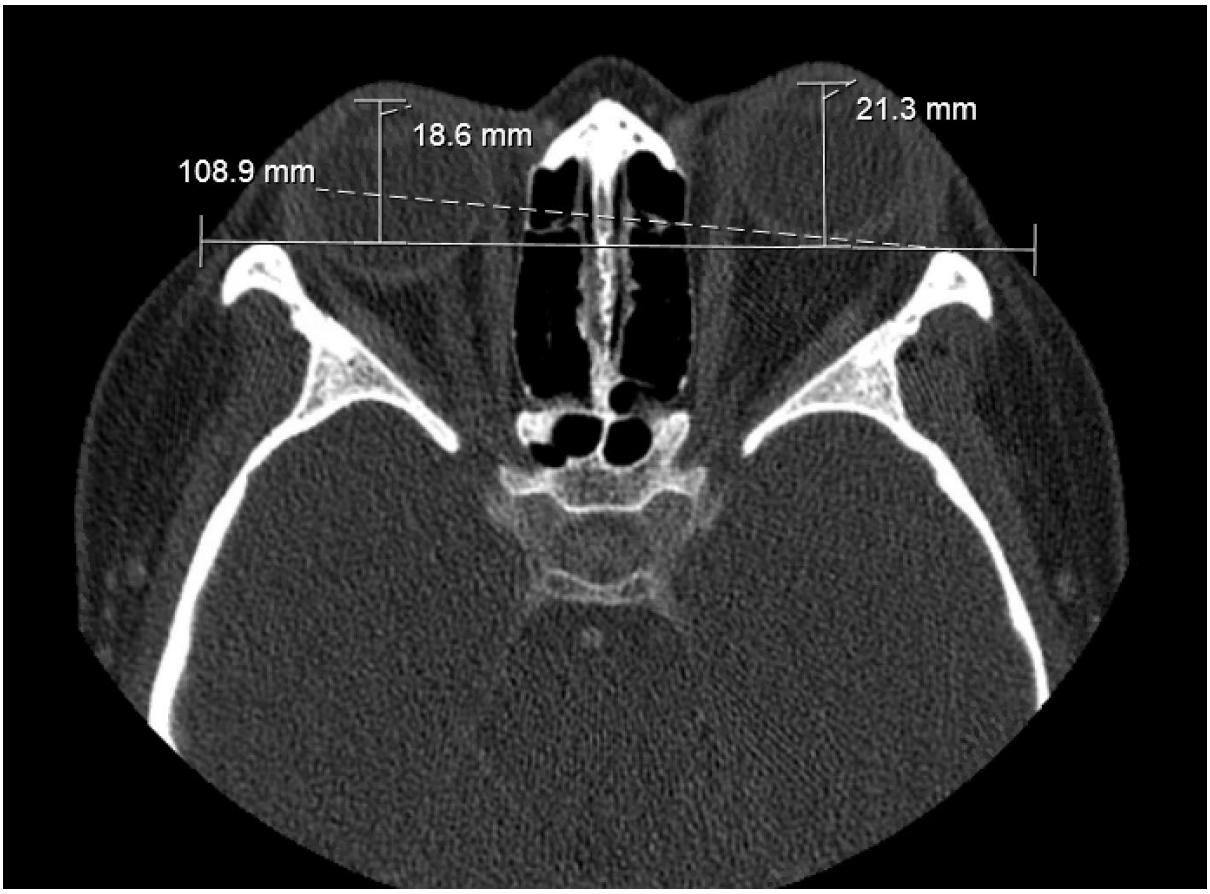

Figure No. 2: CT scan of the head and neck post IV contrast showing complex solid and cystic tumor within the left maxillary sinus, extending into the ipsilateral orbital cavity with resultant ocular proptosis.

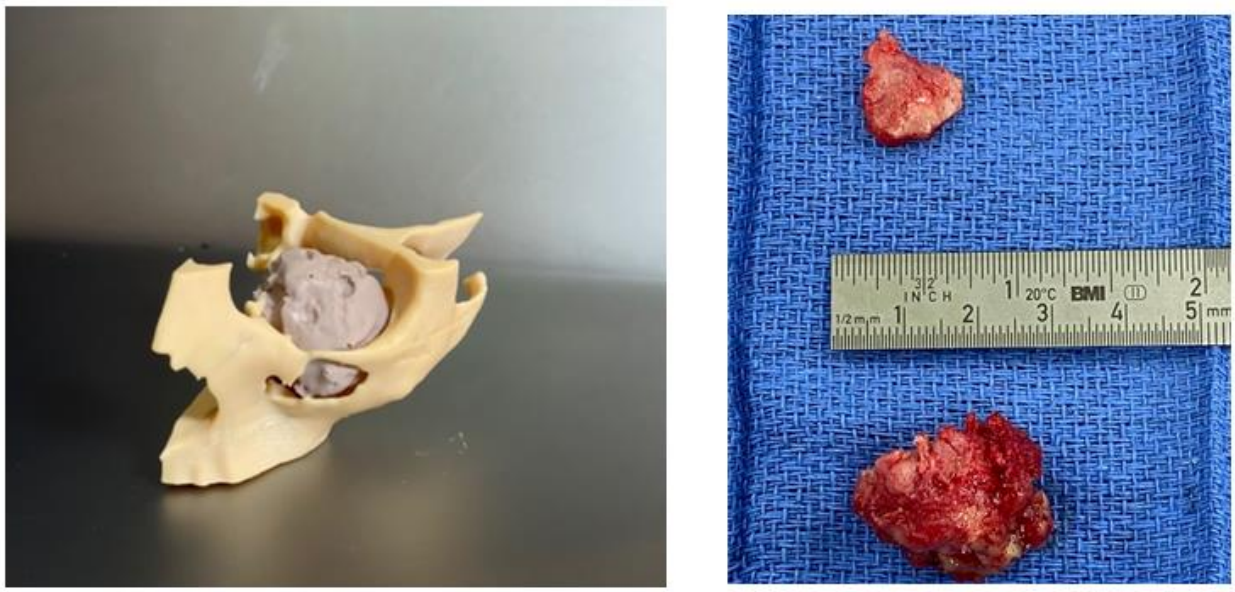

Figure No.3:A 3D printed model showing the lesion to guide the Surgical Procedure (A) picture of the surgical specimen after the excision (B, C)

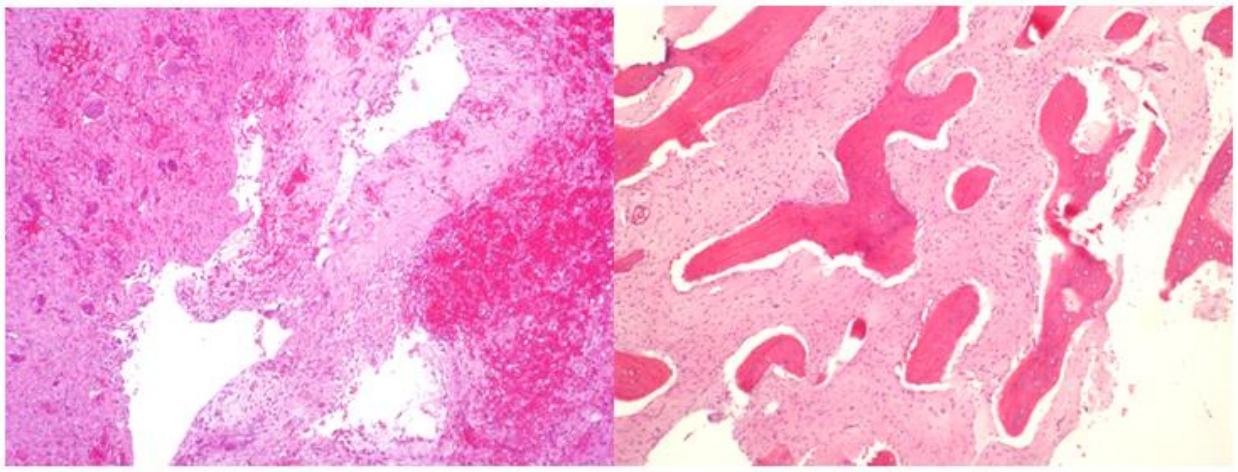

Figure No.4: Histopathology report consistent with juvenile trabecular ossifying fibroma with secondary Aneurysmal bone cyst

DISCUSSION

Ossifying fibroma (OF) is a neoplasm characterized by the formation of fibro-osseous tissue in the craniofacial skeleton. It is a benign tumor that grows slowly but can exhibit local aggressiveness. A notable feature of OF is its well-defined borders, allowing for potential enucleation of the tumor in certain cases. Alongside other benign neoplasms like fibrous dysplasia and osteoma, OF belongs to the broader category of benign fibro-osseous tumors in the craniofacial region. OF itself encompasses various subtypes based on histopathological characteristics, including juvenile trabecular ossifying fibroma, juvenile psammomatoid ossifying fibroma, and cemento-ossifying fibroma.[5]

The exact cause of OF remains unknown, although several possible etiologies have been proposed. Trauma was suggested as a potential origin as early as 1983, but its role is challenged by the absence of facial trauma or orthodontic procedures in our patient's case. Another recently proposed etiology is a genetic factor, as genetic mutations happening in the HRPT2 gene have been identified in sporadic cases, indicating a potential link.[6]

OF typically arises in the mandible or maxilla, but it can also occur outside the jaw in the nose and paranasal sinuses. The ethmoidal sinus is the most common extra-jaw location, although maxillary OF tends to exhibit greater aggressiveness. The age of presentation for OF in the nasal cavity and paranasal sinuses varies widely, with the most commonly encountered age group being in the second decade, with an average age of 23.

OF is often detected incidentally during imaging studies conducted for unrelated reasons due to its painless and gradual growth pattern, as well as its benign nature. When symptoms do occur, they are typically associated with the mass effect caused by the tumor. Common symptoms include facial swelling, headaches, proptosis (protrusion of the eyeball), nasal obstruction, and visual disturbances. Less frequently, patients may experience rhinorrhea (runny nose), recurrent sinusitis, or epistaxis (nosebleeds). Computed Tomography (CT) scans reveal well-defined, initially radiolucent masses that become radiopaque over time. The central region of these masses exhibits a heterogeneous matrix characterized by diffuse calcification, fibrous tissue, or retained mucus, resulting in a ground glass opacification appearance on CT scans.[7]

OF is diagnosed through histopathological examination, necessitating a biopsy of the lesions. Histologically, OF is characterized by a collagenous fibroblastic stroma containing bone structures, including woven bone, lamellar bone, and mineralized components. While these features are commonly observed in OFs, their proportions can vary between different types and individual cases. ABCs are typically benign and well-defined lesions that can also exhibit expansile and destructive properties. They are characterized by the presence of multiple chambers separated by fibrous tissue septae of varying thicknesses. These chambers are often filled with fresh blood or clots, contributing to the characteristic appearance of ABCs.

ABCs primarily affect long bones, with only a small percentage (1.9%) occurring in the head and neck region. Among ABCs in the head and neck, approximately 66% are found in the mandible, while cases of ABCs in the paranasal sinuses are exceptionally rare. ABCs in the sinonasal tract do not show a specific preference for age or gender. The etiology of ABCs can be classified into primary lesions or secondary lesions associated with an underlying neoplasm. Primary lesions are believed to arise de novo and are characterized by a rearrangement in the USP6 gene, resulting in a fusion between the USP6 gene and the CDH11 gene. However, secondary lesions in the head and neck region are primarily caused by conditions such as giant cell granulomas, ossifying fibromas, or fibrous dysplasia and do not exhibit the aforementioned genetic mutation.[8]

ABCs commonly present without pain and can exhibit a wide range of clinical features, ranging from asymptomatic to symptoms such as anosmia, proptosis, or even neurological manifestations. Radiologically, ABCs are observed as multiloculated cysts with expanding borders and lytic characteristics on X-ray. CT and MRI scans provide more detailed information, revealing septae of fibrous tissue that typically run perpendicular to the cortex and can be further enhanced with contrast. Additionally, the cysts themselves display a fluid-fluid level within, which can be visualized using either CT or MRI imaging techniques.[9] The clinical features and radiological findings associated with ABCs are not sufficient for a definitive diagnosis, highlighting the need for a biopsy of the lesion and subsequent histopathological examination. Contrary to its name, an ABC is not lined with endothelium but instead with fibrous septae containing osteoclast-like giant cells, which are relatively specific to ABC. Focal osteoid formations may also be present alongside the fibrous septae and giant cells. The potential causative relationship between OF and ABCs has been suggested, as secondary ABCs often accompany other neoplasms; however, further research is required to establish this relationship more definitively (ABC treat). In our investigation of cases involving OF and ABC in the MS, we identified only five cases, with the earliest report dating back to 1978 and the remaining cases being more recent.[10]

The primary approach to managing both OF and ABCs is surgical resection.[11] However, alternative techniques have also been proposed. It has been suggested that ABCs can be effectively treated with biopsy or curettage alone, as mentioned in the 19th citation of ABC treatment.[12] In specific cases where surgical intervention is not feasible, open packing may be considered a viable option. Regarding OFs, there is no definitive consensus on the optimal treatment approach, but options can range from surgical excision to complete resection.

CONCLUSION

This current case report concludes with clinical importance, appropriate investigation prior to and post-surgical, and the importance of interdisciplinary approaches in the timely management of the above case with the best outcome of the procedure.

Footnotes: Patient consent: Obtained

Author's Contribution:

Concept & Design of Study:	Khalid Zabin Alotaibi
Drafting:	Khalid Zabin Alotaibi
Data Analysis:	Khalid Zabin Alotaibi
Revisiting Critically:	Khalid Zabin Alotaibi
Final Approval of version:	Khalid Zabin Alotaibi

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REFERENCES

1. MacDonald-Jankowski DS. Ossifying fibroma: a systematic review. Dentomaxillofacial Radiol 2009;38(8):495-513.
2. Yadav R, Gulati A. Peripheral ossifying fibroma: a case report. J Oral Sci 2009;51(1):151-4.
3. Farquhar T, MacLellan J, Dyment H, Anderson RD. Peripheral ossifying fibroma: a case report. J Can Dent Assoc 2008;74(9):809-12.
4. Liu Y, Wang H, You M, Yang Z, Miao J, Shimizutani K, Koseki T. Ossifying fibromas of the jaw bone: 20 cases. Dentomaxillofacial Radiol 2010;39(1):57-63.
5. Moon WJ, Choi SY, Chung EC, Kwon KH, Chae SW. Peripheral ossifying fibroma in the oral cavity: CT and MR findings. Dentomaxillofacial Radiol 2007;36(3):180-2.
6. Chaudhari S, Umarji HR. Peripheral ossifying fibroma in the oral cavity: MRI findings. Case Reports in Dentistry 2011;Oct 20.
7. Asaumi JI, Konouchi H, Hisatomi M, Matsuzaki H, Shigehara H, Honda Y, Kishi K. MR features of aneurysmal bone cyst of the mandible and characteristics distinguishing it from other lesions. Eur J Radiol 2003;45(2):108-12.
8. Nasser MJ. Psammomatoid ossifying fibroma with secondary aneurysmal bone cyst of frontal sinus. Child's Nervous System 2009;25:1513-6.
9. Urs AB, Augustine J, Arora S, Kumar P. Rare pediatric presentation of aneurysmal bone cyst with trabecular juvenile ossifying fibroma and ossifying fibroma. Int J Pediatr Otorhinolaryngol 2013; 77(4):576-80.
10. Yazici B, Yazici Z, Yalçinkaya U. Aneurysmal bone cyst secondary to ossifying fibroma in the orbit. Ophthalmic Plastic Reconstructive Surg 2011;27(4):e84-5.
11. Saad R, Lutz JC, Riehm S, Marcellin L, Gros CI, Bornert F. Conservative management of an atypical intra-sinusal ossifying fibroma associated to an aneurysmal bone cyst. J Stomatol Oral Maxillofacial Surg 2018;119(2):140-4.
12. Tamgadge S, Avinash T, Bhalerao S, Rajhans S. Juvenile psammomatoid ossifying fibroma with aneurysmal bone cyst in the posterior mandible. ecancermedicalsci 2014;8.