

*Case Report*

## **Psammomatoid Ossifying Fibroma of the Middle Turbinate with Secondary aneurysmal Bone Cyst**

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### **ABSTRACT**

Ossifying fibroma is a true heterogeneous neoplasm predominantly involving craniofacial bones. Ossifying fibroma of the middle turbinate is extremely rare. There is an infrequent occurrence of a secondary aneurysmal bone cyst in facial bones. Here we report a case of a 24 year old male with an Ossifying fibroma of the middle turbinate associated with a secondary aneurysmal bone cyst diagnosed after a clinicopathological and radiologic correlation. To our knowledge this is the first case involving whole of the middle turbinate with a secondary aneurysmal bone cyst.

**Key words:** Psammomatoid ossifying fibroma; Aneurysmal bone cyst; middle turbinate.

### **INTRODUCTION**

Ossifying fibroma (OF) is a benign fibro-osseous tumour which has an infrequent occurrence in the sinonasal area. It is composed of bone, fibrous tissue and cementum. First described in 1872 by Menzel and the term ossifying fibroma was coined by Montgomery in 1927. <sup>(1)</sup> It has been reported in the literature since 1938 with different names until the WHO classified it as a separate entity in 1992. <sup>(2)</sup> It is predominantly a lesion of the gnathic area. These are radio logically well-defined single bony expansile tumours <sup>(3)</sup> with progressive proliferative capability. There are two variants of the tumour i.e. Psammomatoid and Trabecular variants. There is an infrequent occurrence of aneurysmal bone cyst (ABC) along with ossifying fibroma. <sup>(4)</sup> Aggressive growth is seen when there is a secondary aneurysmal bone cyst. Identified radiologically and a definitive diagnosis is made only histopathologically with clinical correlation.

We report a case of ossifying fibroma of middle turbinate with accompanying aneurysmal bone cyst in a 24 year old male because of its clinical rarity.

### **CASE REPORT**

A 24 year old male presented with chief complaints of right nasal obstruction since two and half years, foul smelling, non-blood stained discharge from right nostril and anosmia on right side since 6 months. He also complained of post nasal drip, epiphora and blurring of vision in his right eye. There was no history of any trauma, headache, double vision, fever, retro orbital pain, tooth ache, weight loss and loss of appetite.

On Examination, dorsum of the nose was pushed to the left. There was fullness present at the medial canthus of right eye with an increase in the right intercanthal distance Figure 1. Nasal endoscopy showed a firm mucosa lined mass completely obstructing the right nasal cavity which did not bleed on touch. Nasal septum was

pushed to the left. Gingivo labial sulcus and hard palate were within normal limits. Eye movements were normal although blurring of vision was present.

A contrast enhanced computed tomography showed an expansile 5x5.2x5.3cm mass with calcifications and hypodense water attenuation arising from the middle turbinate obliterating right nasal cavity completely with minimal contrast enhancement. Periphery of the lesion showed marked bony remodeling extending into the right frontal and maxillary sinuses. Nasal septum and the medial wall of the right orbit were thinned out and bowing towards the left and the orbital surface respectively. Figure 2



Figure 1: Showing increased right intercanthal distance



Figure 2: Contrast enhanced Computed Tomography

Based on a provisional diagnosis of ossifying fibroma endoscopic resection was done. Frank bleeding from the mass was seen on puncturing the outer wall. On breaking the outer wall, trabeculae and fibrous septae were seen which were broken

and complete resection of the tumour was done in piecemeal and sent for histopathology Figure 3 Figure 4.

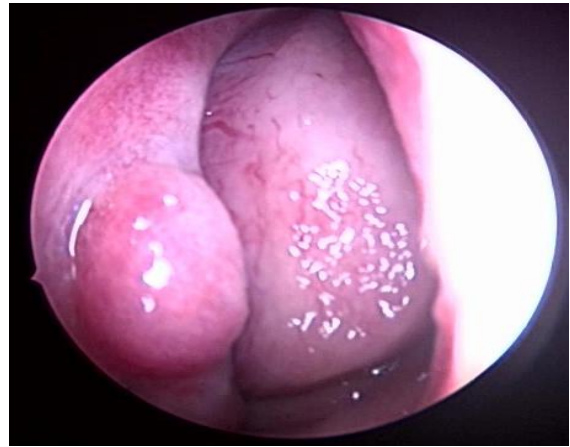


Figure 3: DNE showing a firm mass completely obstructing the nasal cavity.

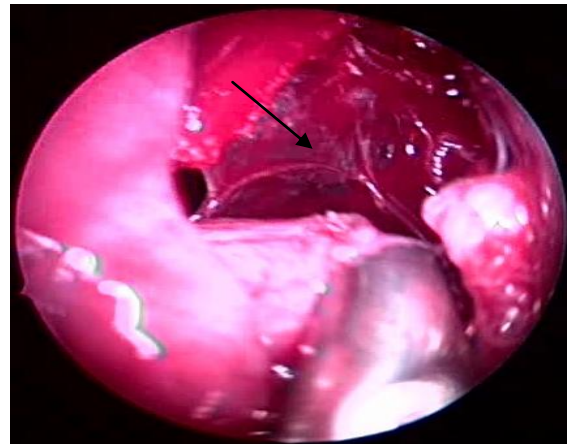


Figure 4: Trabeculae with fibrous septa seen after removing the outer shell

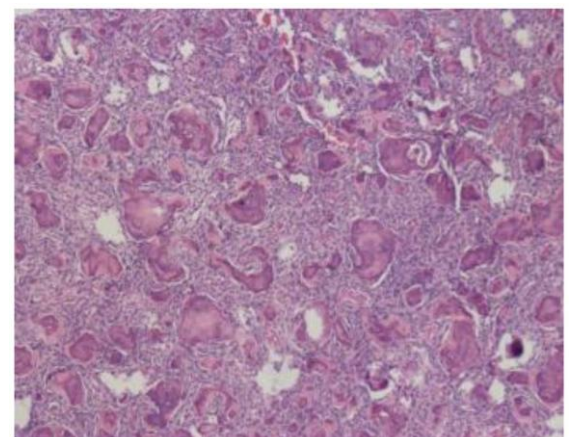


Figure 5: H & E staining with Characteristic Psammomatoid bodies.

Histology showed pathognomonic Psammomatoid calcifications with hypercellular stromal elements containing interspersed with vascular spaces containing

giant cells diagnostic Figure 5 of psammomatoid ossifying fibroma of the middle turbinate with secondary aneurysmal bone cyst. At one year of follow up there is no evidence of recurrence.

## DISCUSSION

Ossifying fibroma is a true heterogeneous<sup>(4)</sup> neoplasm with variable presentation depending on the site of involvement. It predominantly affects the craniofacial bones with mandible at the top of the list with 77%.<sup>(1)</sup>

A secondary lesion like aneurysmal bone cyst is not uncommon in a fibro-osseous lesion. Less than 5% of ABC has been found in craniofacial bones. They typically occur in long bones. Their occurrence in nasal cavity and a paranasal sinus is very rare. They are expansile multilocular, multicystic lesions having fibrous septae and reactive bony trabeculae containing extravasated RBCs and chronic inflammatory infiltrates.

Predominantly there are 2 types of ossifying fibroma, Trabecular (Tr JOF) and Psammomatoid (Ps JOF) based on their histological appearance.<sup>(5)</sup> They are more common in children and young adults with an average age of 8-12 yrs for Tr JOF and 16-33 yrs for Ps JOF.<sup>(4)</sup> Occurrence of secondary aneurysmal bone cyst is frequently associated with psammomatoid type than the trabecular.<sup>(15-17)</sup> Trabecular type involves the jaw bones whereas the psammomatoid type in the periorbital, frontal and ethmoid bones. Ossifying fibroma of the middle turbinate is extremely rare, where the first case was reported by Caylakli et al in a 28 yr old female only in 2004.<sup>(6)</sup> Since then only four other cases of ossifying fibroma of the middle turbinate has been documented.<sup>(7-10)</sup> Nevertheless here we have a first case of ossifying fibroma of the middle turbinate with a secondary aneurysmal bone cyst.

Psammomatoid OF are reported 4 times more than the trabecular variant. Aggressive growth is seen in younger patients and when they are associated with

ABC. They have to be differentiated from other fibro-osseous lesions like fibrous dysplasia which usually does not progress after puberty. They invade the adjacent structures like the orbit, skull base where they present with loco regional symptoms like proptosis, epiphora, exophthalmos and diplopia.<sup>(14)</sup>

OF is hypothesized to be originating either from the periodontal membrane which migrated to other areas during embryogenesis or the periodontal ligament in the tooth bearing areas of jaw capable of differentiating to cementum, bone and fibrous tissue.<sup>(11)</sup>

Pathogenesis of secondary aneurysmal bone cyst is still debatable. There are three theories which might explain the formation of ABC in OF. First, due to overproduction of the myxofibrous cellular stroma involved in development of septae in the paranasal sinuses. These stromal cells secrete hyaline material that ossifies and connective tissue material that forms cystic spaces.<sup>(12)</sup> Second, intrabony vascular defect predisposed by the extensive vascularity and the consequent change in the hemodynamics.<sup>(13)</sup> Third, cell edema of ossifying fibroma may easily cause microcyst formation which may further develop into ABC. Aggressive growth is seen when associated with an aneurysmal bone cyst.<sup>(4)</sup> Contrast enhanced CT shows round oval heterogeneous mass, expansile with thin bony egg-shelling interspersed with calcified areas with peripheral bony remodeling. Attenuation ranges from radiolucent to ground glass to sclerotic changes. Multicystic appearance with thin septae showing water like low attenuation is suggestive of a secondary aneurysmal bone cyst.<sup>(15)</sup> MRI shows isointense to grey matter T1WI and hypo-isointense T2WI with moderate to marked contrast enhancement. Fluid-Fluid levels can be seen in T2WI showing upper hyperintense and lower hypo-isointense part diagnostic of aneurysmal bone cyst. However in our case due to the low socio economic status of the patient MRI could not be done.

Presence of round psammomatoid bodies in a hypercellular fibrous stroma along with vascular spaces lined by giant cells filled with RBC's and inflammatory cells are characteristically diagnostic of ossifying fibroma with an aneurysmal bone cyst. (7)

Complete resection of the tumour is the treatment of choice. An endoscopic resection can be done depending on the extent of the disease and surgical expertise as has been performed in the present case. Radiation is contraindicated due to the risk of malignant transformation.

Recurrence has been reported in the range of 30-58%. Incomplete resection is associated with residual disease and recurrences. (4)

## CONCLUSION

Psammomatoid Ossifying fibroma is a unique Fibro osseous lesion of the sinonasal region well demarcated expansile multiloculated swelling with cortical thinning. It is aggressive in children, young adults and when associated with an aneurysmal bone cyst. (4)

A Clinical, Radiological and Histopathological correlation is needed for a definitive diagnosis. (9,14,17) Complete excision with long term follow up is needed to manage the recurrences.

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