Sinonasal Ossifying Fibroma: A Study of Six Cases and Review of Literature
B Vikram, S Udayashankar

Citation

Abstract
Background: Ossifying fibroma is an uncommon tumor of the craniofacial skeleton presenting in several variant histopathological subtypes. The overlapping clinical and histopathological features of these subtypes have led to diagnostic dilemma and confusion. Complete excision of this tumor has become a necessity since it is notorious for recurrence.

Aims: To study and compare the clinical profiles of various types of ossifying fibroma and also the surgical approaches to the tumor. To discuss the diagnostic difficulties and controversies associated with the tumor.

Setting: Medical college referral hospital.

Design: Prospective descriptive study of 2 years duration.

Methods: Planned surgical excision based on criteria. Literature review with Medline search.

Results: Six cases of different types of ossifying fibroma were successfully treated by various surgical approaches. Thirty papers related to diagnostic and treatment aspects of ossifying fibroma were studied. Eight papers were found to be associated with some or the other controversy.

Conclusion: The diagnostic dilemma of ossifying fibroma can be overcome with a combination of clinical, radiological and pathological criteria. Complete surgical excision of the tumor is possible when surgery is based on preplanned criteria. Combination of 2 or more surgical approaches may be necessary in many cases in order to ensure complete clearance and prevent recurrence of the tumor. To avoid controversies, there should be a consensus on nomenclature and classification of this uncommon tumor.

KEY MESSAGES
1. Diagnostic dilemma of ossifying fibroma can be overcome with a combination of clinical, radiological and pathological criteria.
2. It is preferable to remove the tumor enmass, take multiple sections for histopathological reporting to avoid missing the subtypes.
3. Complete surgical excision of the tumor is possible when surgery is based on preplanned criteria.

INTRODUCTION
Ossifying fibroma is a destructive, deforming, slow growing, benign fibro-osseous tumor that can occur almost anywhere in the facial skeleton. This uncommon tumor can present a diagnostic dilemma for the clinician and the pathologist, owing to overlapping clinical and histomorphologic features. Many synonymous nomenclatures exist for a single entity and the controversy in classification and staging of the subtypes in the literature has added to the confusion. Hence there is a need to highlight the points of controversy existing for this tumor so that they could be avoided through a consensus in future. The tumor can produce sinus obstruction, infection, facial deformity, proptosis and intracranial complications, even though it can remain asymptomatic in the early stage. Therefore the tumor needs to be excised completely in order to prevent recurrence.
Complete excision is dependent on the correct surgical approach. Hence we decided to base our surgeries on preplanned criteria to excise the tumor completely and minimize recurrence. This paper has been presented with the following objectives:

1. To present 6 cases of ossifying fibroma that were successfully treated by different surgical approaches by us
2. To compare the clinical profiles of various types of ossifying fibroma and discuss the controversies those exist for this tumor in literature.
3. To discuss the diagnostic dilemma associated with ossifying fibroma
4. To discuss the various surgical approaches to this uncommon tumor in the sinonasal region.

**MATERIALS AND METHODS**

This is a prospective descriptive study of all cases of sinonasal ossifying fibroma that were diagnosed and treated in a period of 2 years in a medical college referral hospital catering to both urban and rural population. Cases from all ages and both sexes were included. Cases with tumor involving the orbit and the cranial cavity were also included. But those cases with the tumor involving the lower jaw, recurrences of tumor and other conditions like fibrous dysplasia were excluded. The diagnostic details of the patients are described in table 1.

**Figure 1**

![CT Scan of the head (Axial section case 1)](image)

**Table 1**: Diagnostic details of the individual cases

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Tumor mass</th>
<th>Examination</th>
<th>Radiographic diagnosis involved</th>
<th>Diagnostic nasal endoscopy</th>
<th>Biopsy report</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>25</td>
<td>F</td>
<td>orbital</td>
<td>CT scan</td>
<td>Mass in right orbit</td>
<td>No change in bone density</td>
<td>Normal</td>
</tr>
<tr>
<td>2</td>
<td>35</td>
<td>M</td>
<td>nasal cavity</td>
<td>CT scan</td>
<td>Mass in right nasal cavity</td>
<td>No change in bone density</td>
<td>Normal</td>
</tr>
<tr>
<td>3</td>
<td>40</td>
<td>M</td>
<td>nasal cavity</td>
<td>CT scan</td>
<td>Mass in left nasal cavity</td>
<td>No change in bone density</td>
<td>Normal</td>
</tr>
<tr>
<td>4</td>
<td>50</td>
<td>F</td>
<td>nasal cavity</td>
<td>CT scan</td>
<td>Mass in nasal cavity - thin, spongy</td>
<td>No change in bone density</td>
<td>Normal</td>
</tr>
<tr>
<td>5</td>
<td>60</td>
<td>M</td>
<td>nasal cavity</td>
<td>CT scan</td>
<td>Mass in nasal cavity - thin, spongy</td>
<td>No change in bone density</td>
<td>Normal</td>
</tr>
<tr>
<td>6</td>
<td>70</td>
<td>F</td>
<td>nasal cavity</td>
<td>CT scan</td>
<td>Mass in nasal cavity - thin, spongy</td>
<td>No change in bone density</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Investigations such as X-ray paranasal sinuses [PNS], CT scan PNS (Fig 1 to 3), diagnostic nasal endoscopy (Fig 4) and biopsy were performed on all the patients. The classical ‘punched out appearance’ and ‘egg shell expansion’ of the tumor can be appreciated in the CT scan of case 2 (Fig 2).

**Figure 2**

Figure 1: CT Scan of the head (Axial section case 1) showing tumor mass in the medial part of the right orbit.

**Figure 3**

Figure 2: CT Scan of the head (Coronal section case 2) demonstrating ‘punched out appearance’ and ‘egg shell expansion’ of the tumor.
We decided to plan our surgeries based on the following criteria:

1. To perform the least radical tumor resection possible
2. To excise the tumor in toto and also try to remove it enmass wherever possible.
3. To cause least damage to the surrounding structures that might affect the growth of the craniofacial skeleton
4. To try to avoid an external scar, especially in pediatric patients
5. To try to prevent recurrence of tumor

**SURGICAL APPROACHES**

The type of approach was decided by the following factors:

1. Age of the patient
2. Presence of complications
3. Histopathological type of tumor
4. Surgical criteria already mentioned

A Medline literature search with key words ‘ossifying fibroma’ was made to look for the references on the nomenclature, classification, diagnostic details and treatment of ossifying fibroma.

**RESULTS**

Six cases of craniofacial ossifying fibroma were surgically treated by various surgical approaches. All the 6 patients were operated under general anesthesia. Details of the surgeries are given in table 2. The photograph of the tumor mass that was excised in case 2 is shown in figure 5. In case 4, a dual approach had to be employed due to the significant intracranial extension of the tumor. The help of a neurosurgeon was taken to excise the intracranial portion of the tumor by a bifrontal craniotomy. The CSF leak was sealed and the dura was repaired with fascia lata. The nasal part of the tumor was removed piecemeal by rigid nasal endoscopy in the same sitting. Intraoperative bleeding was moderate in all cases except in case 5 where blood transfusion had to be given. The excised mass in all cases was sent for histopathological examination. An endoscopic assessment of the surgical cavity after tumor excision was
done in every case. No recurrence was observed clinically, endoscopically and radiologically in any of the cases in a follow up period of one year.

**Figure 7**

Table 2: Surgical details of the individual cases

<table>
<thead>
<tr>
<th>Surgical site code</th>
<th>Surgical site region</th>
<th>Pathological diagnosis</th>
<th>Follow up</th>
<th>Post surgery side effects</th>
<th>Duration of surgery (minutes)</th>
<th>Complications</th>
<th>Follow up period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>Lynch syndrome, extraoral</td>
<td>Enameloid odontogenic fibroma, cystic</td>
<td>Good, 7 days&lt;br&gt;Medial to lateral</td>
<td>Nil</td>
<td>Nil</td>
<td>No recurrence</td>
<td>12 months</td>
</tr>
<tr>
<td>Case 2</td>
<td>Lynch syndrome, intraoral, nasal</td>
<td>Enameloid odontogenic fibroma, cystic</td>
<td>Good, 10 days&lt;br&gt;Medial to lateral</td>
<td>Minor surgical complications</td>
<td>Nil</td>
<td>No recurrence</td>
<td>12 months</td>
</tr>
<tr>
<td>Case 3</td>
<td>Partial calvarial, frontal</td>
<td>Enameloid odontogenic fibroma, cystic</td>
<td>Good, 9 days&lt;br&gt;Mild to moderate</td>
<td>Minor surgical complications</td>
<td>Nil</td>
<td>No recurrence</td>
<td>12 months</td>
</tr>
<tr>
<td>Case 4</td>
<td>Enameloid odontogenic fibroma, maxillary sinuses</td>
<td>Enameloid odontogenic fibroma, cystic</td>
<td>Good, 10 days&lt;br&gt;Medial to lateral</td>
<td>Minor surgical complications</td>
<td>Nil</td>
<td>No recurrence</td>
<td>12 months</td>
</tr>
<tr>
<td>Case 5</td>
<td>Enameloid odontogenic fibroma, maxillary sinuses</td>
<td>Enameloid odontogenic fibroma, cystic</td>
<td>Good, 9 days&lt;br&gt;Mild to moderate</td>
<td>Minor surgical complications</td>
<td>Nil</td>
<td>No recurrence</td>
<td>12 months</td>
</tr>
<tr>
<td>Case 6</td>
<td>Enameloid odontogenic fibroma, maxillary sinuses</td>
<td>Enameloid odontogenic fibroma, cystic</td>
<td>Good, 11 days&lt;br&gt;Medial to lateral</td>
<td>Minor surgical complications</td>
<td>Nil</td>
<td>No recurrence</td>
<td>12 months</td>
</tr>
</tbody>
</table>

Thirty papers related to the diagnostic and treatment aspects of ossifying fibroma were studied after the Medline search. Eight papers were found to be associated with some or the other controversy. A detailed discussion of the diagnostic difficulties and controversies associated with ossifying fibroma has been made in the discussion part of this paper.

**DISCUSSION**

Ossifying fibroma is a relatively uncommon benign fibro-osseous tumor commonly affecting the craniofacial region. The neoplasm closely resembles the non-neoplastic condition fibrous dysplasia in many respects especially in histology. Hence many pathologists still consider it to be a variant of fibrous dysplasia. The classification of ossifying fibroma with many synonymous nomenclatures has been a matter of controversy. The various names associated with ossifying fibroma are cementifying or cemento-ossifying fibroma, peripheral ossifying fibroma, psammomatoid or juvenile ossifying fibroma and ossifying fibromyxoid tumor. While some authors believe that all these names refer to the same tumor or its variants, others believe that they are separate entities. The absence of typical histopathological features unique to ossifying fibroma and its subtypes or variants has been one of the main reasons for the controversies in reporting. Hybrid fibro-osseous lesions have also been described that contained entities of aneurysmal bone cyst, ossifying fibroma and cementifying fibroma within the same tumor mass. This suggests that simultaneous occurrence of multiple related lesions is possible in the same tumor mass, which can further complicate the overall picture. Hence multiple sections have to be taken for proper histopathological reporting.

The conventional ossifying fibroma usually presents as a solitary, slow growing, monostotic tumor in the third and the fourth decades of life. It shows female predilection and the male to female ratio is around 1:5. Although found predominantly in the mandible (75%), it can also arise in the skull base and PNS. It has also been reported to occur in the temporal bone. The tumor is known to be more aggressive in young patients. In the jaws, the tumor shows affinity for the molar area. In the mandible it can give rise to pathological fractures and osteomyelitis in the long run if left untreated.

Psammomatoid or juvenile ossifying fibroma is an aggressive variant of ossifying fibroma usually seen in the pediatric age group that is histologically characterized by the formation of Psammoma body islands. Juvenile ossifying fibroma has been further classified into 2 separate subtypes as psammomatoid and trabecular ossifying fibroma. Psammomatoid ossifying fibroma of the paranasal sinuses has been considered as an extragnathic variant of cemento-ossifying fibroma by some authors. Some authors consider juvenile active ossifying fibroma and Psammomatoid ossifying fibroma as separate entities. Juvenile or psammomatoid ossifying fibroma has its peak incidence in the I and II decades of life with a female predilection. Twenty one percent of the patients are older than 15 years and hence some clinicians refuse to accept the word ‘juvenile’. The preferred sites are the maxilla, ethmoid and the frontal bones. The term ‘active’ refers to clinical aggressive behavior such as bone erosion, soft tissue invasion and in rare instances death. The most common clinical manifestation is proptosis. Other symptoms include nasal obstruction, headache, swelling and rarely epistaxis. The most distinctive component in these lesions (Even though not pathognomonic) is the presence of calcified ossicles containing osteocytes known as ‘psammomatoid bodies’. Because of a superficial resemblance between these ossicles and the cementum spheres of the odontogenic ossifying fibroma, the lesion has sometimes been mislabeled as cemento-ossifying fibroma, implying an odontogenic origin, which is rather unlikely in extragnathic bone. Hence Psammomatoid bodies have been synonymously referred to as ‘cementicles’ by some authors.

Ossifying fibromyxoid tumor is a recently described mesenchymal neoplasm that can also occur in the head and neck region. It was originally defined as a borderline or low-grade malignant lesion. Some authors have considered it as a tumor of intermediate malignancy that can also give rise to
metastases. The tumor has been further classified into ‘typical’, ‘atypical’ and ‘malignant’ subtypes. The tumor usually occurs in the deep subcutis of the extremities, trunk and the head and neck region. Histologically the tumor is characterized by the presence of a peripheral rim of lamellar bone within the collagogenous capsule, a lacelike and glomoid arrangement of tumor cells in a fibromyxoid background and also round tumor cells that have a bland cytologic appearance. The histogenesis of this tumor is uncertain although the preponderance of evidence suggests a Schwann cell origin. 

Cementifying fibroma is a distinctive jaw lesion and has its origin from the periodontal membrane that has the characteristic feature of cementum formation. Hence it is also known as periodontoma. The term cementifying fibroma has been applied to lesions containing curvilinear trabeculae and spheroidal calcifications (Cementum). Cementifying fibroma most frequently occurs in the III and IV decades primarily affects females and typically involves the mandibular premolar-molar area. Some authors consider cemento-ossifying fibroma separately and refer them to lesions containing bone and cemental tissues. These lesions are usually found in the tooth bearing areas of the jaws. Pediatric dentists have referred to a reactive gingival nodule arising from the periodontal ligament as peripheral ossifying fibroma that is probably similar to cementifying fibroma.

Interestingly, some authors have suggested that all the fibro-osseous lesions relating to ossifying fibroma and its subtypes should be referred to as ossifying fibroma only since they claim that there is no difference in behavior between the subtypes and the histological designations are only academic. Even though some authors opine that fibrous dysplasia and ossifying fibroma cannot be distinguished by microscopy due to histological overlap, others opine that there are many features like the absence of cementicles, lamellar trabeculae and osteoblasts to distinguish them. The differentiation between the 2 entities is of great importance because of their divergent clinical behavior. Fibrous dysplasia is often polyostotic whereas ossifying fibroma is usually monostotic. Fibrous dysplasia is usually a self-limiting disease and therefore a complete resection is unnecessary and in most instances impossible. Radiographically ossifying fibromas are classically described as circumscribed unilocular lesions with a surrounding rim of eggshell thin bone giving a ‘punched out

'appearance. Irregular thickening and thinning of the edges of this eggshell has been described as ‘moth-eaten’ appearance. In contrast, fibrous dysplasia has a ‘ground glass’ appearance with indistinct borders that blend imperceptibly into the adjacent bone without expansion or bone destruction. But again some authors have also described ‘ground glass’ appearance in juvenile active ossifying fibroma. Hence CT scan with the bone window technique is the most essential investigation since it defines the extent of the tumor and the destruction of the surrounding tissues. The differential diagnosis of ossifying fibroma includes fibrous dysplasia, giant cell tumor, giant cell reparative granuloma, aneurysmal bone cyst, osteoblastoma and osteosarcoma.

The overall prognosis with most types of ossifying fibroma appears to be good. Despite their tendency for local invasion and recurrence, there are no reported instances of metastatic disease with the exception of certain subtypes of ossifying fibromyxoid tumor. The development of aneurysmal bone cyst in psammomatoid ossifying fibroma has been reported. Malignant transformation is very rare. Meningitis secondary to invasion into the cranial cavity has been reported and rarely even death may occur. Surgery is the mainstay of treatment for all types of ossifying fibroma. Growth of the facial skeleton is an important consideration in the pediatric age group. Extensive tumors in this age group are more difficult to manage since the tumors here tend to be more invasive, aggressive and have more chances of recurrence. Mandibular tumors can be treated with conservative surgery but aggressive surgery is warranted for midface and paranasal sinuses because of their more aggressive behavior. Even though small tumors can be removed by simple curettage and enucleation, they are better avoided for the fear of recurrence especially in juvenile ossifying fibroma. One study showed a recurrence rate of 28% after curettage. Endoscopic removal has also been tried successfully. But these conservative techniques and piecemeal approaches could make histological interpretation more difficult, especially in cases of hybrid lesions. Hence wherever possible, an open surgical technique is advised for adequate visualization and complete excision. Various radical and semi radical resection procedures have been tried. Fronto-parietal craniotomy with cranioplasty has been done for lesions of the parietal bone. Similarly partial mandibulectomy followed by plate fixation has been done for mandibular lesions. Extensive bilateral tumors require combined multiple approaches whereby the tumor is excised.
portion by portion. Bilateral nasal tumors have been removed by modified lateral rhinotomy with bilateral ethmoidectomy and sphenoidotomy. Larger lesions with intracranial extension may require en bloc resection or craniofacial resection.

Surgical complications may include significant blood loss, requiring transfusion and loss of vision. Recurrences after surgery have been common and reported to be in the range of 30 to 56%. Treatment of recurrences by revision surgery is difficult again in the pediatric group. Radiotherapy has been proven to be ineffective and is contraindicated due to an increased incidence of malignant transformation ranging from 0.4 to 44% with the exception of certain subtypes of ossifying fibromyxoid tumor.

In our experience, the Lynch Howarth approach has been particularly useful to relieve proptosis when the tumor was confined only to the medial portion of the orbit and the ethmoid. The extended Caldwell Luc approach and sublabial approach are more suitable for lesions involving the maxilla and the premaxilla and had the advantage of having no external scar. Lateral rhinotomy approach gave the widest access and exposure to all the regions, but with an external scar. Nasal endoscopic excision had the advantage of tumor excision under direct vision and had no external scar. Hence it could be used as adjuvant for other approaches especially when the tumor was extensive and had to be removed piecemeal. It was also useful to assess the surgical cavity for bleeding and tumor remnants after the excision of the tumor by any approach. But it had the disadvantage of piecemeal removal, risk of recurrence and excessive time consumption for the procedure especially if it was the sole approach employed for removal of the tumor. Hence it appeared to be more suitable for early tumors especially in the pediatric patients where radical surgeries were not advisable. Craniotomy approach had the maximum hospital stay due to higher morbidity.

This study has highlighted the versatile presentation and the heterogeneous nature of this benign tumor in the sinonasal region. Larger comparative studies of longer duration and follow up might be required to suggest an algorithm that could be useful in the decision making process for this tumor. But since the tumor is uncommon, it is difficult to undertake such prospective studies in a reasonable period of time.

CONCLUSION

Ossifying fibroma is a benign fibro-osseous tumor of the craniofacial region that is diagnosed with a combination of clinical, radiological and pathological criteria. Due to the possibility of the presence of hybrid lesions in this tumor, it is preferable to remove it en mass and take multiple sections for histopathological reporting. This would avoid missing a particular subtype of the tumor that might need a different surgical management. The classification of the subtypes and the nomenclature of this tumor have remained controversial. There is a need for consensus regarding the nomenclature and classification of this tumor. The surgical approaches and techniques have also not been well defined especially in the young patients. Complete surgical excision of the tumor is possible when surgery is based on preplanned criteria. Six cases of different types of sinonasal ossifying fibroma managed successfully by various surgical approaches have been presented.

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CORRESPONDENCE TO

Dr. Vikram Bhat K MS, DNB, MNAMS (ORL – HNS) No: 4-64, Sagri Nole, Kunjibettu Post, Udupi – 576102, Karnataka, India Tel (Res): +918362370579 Cell: +919448658213 Email: entvikram@rediffmail.com

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8. Wenig BM, Vinh TN, Smirniotopoulos JG, Fowler CB,


Author Information

Bhat K. Vikram, MS, DNB, MNAMS (ORL - HNS)
Associate Prof & PhD research scholar in Otolaryngology, Head & Neck Surgery, Karnataka Institute of Medical Sciences

S. G. Udayashankar, MS, DLO
Prof & Head of the department of Otolaryngology, Head & Neck Surgery, Karnataka Institute of Medical Sciences