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Assessment of the fibro-osseous lesions of the paranasal sinuses and the skull base

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Abstract
The vast majority of fibro-osseous lesions are incidental findings on radiographs. They are slow-growing tumors, and are infrequently symptomatic. frontal headache is the most common complaint, followed by drainage and nasal obstruction associated with secondary sinusitis. The present study was planned to assess the symptomatic fibro-osseous lesions of the paranasal sinuses.
The study was planned in the Department of ENT in Nalanda Medical College, Patna from Jan 2018 to July 2018. Total 20 patients diagnosed with the fibro osseous lesions were enrolled in the present study. The information regarding patient demographic characteristics, location of the lesion at presentation, CT scan findings, management approach, details of surgery performed, and outcomes were collected and presented as below.
The data generated from the present study concludes that Diagnostic dilemma of benign fibro-osseous lesions of the paranasal sinuses and skull base can be resolved with a combination of clinical, radiological and pathological criteria. Correct diagnosis of fibro osseous tumors is crucial for adequate therapy as their treatment, prognosis, clinical aggressiveness and outcomes of individual entities vary significantly.

Keywords: Fibro-osseous lesions, Osteoma, Fibrous dysplasia (FD), Ossifying Fibroma (OF), Juvenile active ossifying fibroma (JAOF)

Introduction
An osteoma is a new piece of bone usually growing on another piece of bone, typically the skull. It is a benign tumor. When the bone tumor grows on other bone it is known as "homoplastic osteoma"; when it grows on other tissue it is called "heteroplastic osteoma". Osteoma represents the most common benign neoplasm of the nose and paranasal sinuses. The cause of osteomata is uncertain, but commonly accepted theories propose embryologic, traumatic, or infectious causes. Osteomata are also found in Gardner's syndrome. Larger craniofacial osteomata may cause facial pain, headache, and infection due to obstructed nasofrontal ducts. Often, craniofacial osteoma presents itself through ocular signs and symptoms (such as proptosis) [1].
Fibrous dysplasia [FD] is a disorder where normal bone and marrow is replaced with fibrous tissue, resulting in formation of bone that is weak and prone to expansion. As a result, most complications result from fracture, deformity, functional impairment and pain [2]. Disease occurs along a broad clinical spectrum ranging from asymptomatic, incidental lesions to severe disabling disease. Disease can affect one bone (monostotic) or multiple (polyostotic) and may occur in isolation or in combination with café au lait skin macules and hyperfunctioning endocrinopathies, termed McCune–Albright syndrome. More rarely, fibrous dysplasia may be associated with intramuscular myxomas, termed Mazabraud's syndrome [3]. Fibrous dysplasia is very rare, and there is no known cure. Fibrous dysplasia is not a form of cancer.
Treatment in fibrous dysplasia is mainly palliative, and is focused on managing fractures and preventing deformity. There are no medications capable of altering the disease course. Intravenous bisphosphonates may be helpful for treatment of bone pain, but there is no clear evidence that they strengthen bone lesions or prevent fractures. Surgical techniques that are effective in other disorders, such as bone grafting, curettage, and plates and screws, are frequently ineffective in fibrous dysplasia and should be avoided. Intramedullary rods are generally preferred for management of fractures and deformity in the lower extremities. Progressive scoliosis can generally be managed with standard instrumentation and fusion techniques. Surgical management in the craniofacial skeleton is complicated by frequent post-operative FD regrowth, and should focus on correction of functional deformities. Prophylactic optic nerve decompression increases the risk of vision loss and is contraindicated [4].
Managing endocrinopathies is a critical component of management in FD. All patients with fibrous dysplasia should be evaluated and treated for endocrine diseases associated with McCune–Albright syndrome. In particular untreated growth hormone excess may worsen craniofacial fibrous dysplasia and increase the risk of blindness. Untreated hypophosphatemia increases bone pain and risk of fractures [5].
Peripheral ossifying fibroma “a gingival nodule which is composed of a cellular fibroblastic connective tissue stroma which is associated with the formation of randomly dispersed foci of mineralised products, which consists of bone, cementum-like tissue, or a dystrophic calcification. The lesion is considered part of an ossifying fibroma, but that is usually considered to be a jaw tumor. Because of its overwhelming incidence on the gingiva, the condition is associated with two other diseases, though not because they occur together. Instead, the three are associated with each other because they appear frequently on gingiva: pyogenic granuloma and peripheral giant cell granuloma. Some researchers believe peripheral ossifying fibromas to be

related to pyogenic fibromas and, in some instances, are the result of a pyogenic granuloma which has undergone fibrosis and calcification. The term peripheral ossifying fibroma has been criticized as this lesion is not related to the ossifying fibroma of bone and is not a fibroma. This term is used in America, however in Britain, this lesion would be termed a fibrous epulis containing bone [6].

The color of peripheral ossifying fibromas ranges from red to pink, and is frequently ulcerated [7]. It can be sessile or pedunculated with the size usually being less than 2 cm. Weeks or months may pass by before it is seen and diagnosed. There is a gender difference with 66% of the disease occurring in females. The prevalence of peripheral ossifying fibromas is highest around 10 – 19 years of age. It appears only on the gingiva, more often on the maxilla rather than the mandible, and is frequently found in the area around incisors and canines. The adjacent teeth are usually not affected.

Peripheral ossifying fibromas appear microscopically as a combination of a mineralized product and fibrous proliferation. The mineralized portion may be bone, cementum-like, or dystrophic calcifications. Additionally, highly developed bone or cementum is more likely to be present when the peripheral ossifying fibroma has existed for a longer period of time.

A juvenile active ossifying fibroma is a benign fibro-osseous neoplasm composed of mixture of stroma and bone characterized by rapid and destructive growth. This tumor has gone by several names in the past, but active ossifying fibroma is similar to juvenile active ossifying fibroma, except it does not develop in young patients. Aggressive psammomatoid ossifying fibroma is still employed by some, but is to be discouraged. Most patients are asymptomatic, and come to clinical attention when a mass is discovered incidentally on routine dental X-rays. When patients are symptomatic, they present with non-specific symptoms, such as chronic sinusitis, rhinorrhea, obstruction, pain, facial enlargement and possibly visual changes. When performing imaging studies, bone windows in computed tomography studies are the best. The lesion is usually identified as a well demarcated, expansile mass with an ossified rim at the periphery. Calcifications are noted throughout. MRI shows a variable finding depending on T1 or T2 weighted images, dependent on the amount of bone to fibrous connective tissue ratio. The tumors are described as "shelling out" by the surgeon, which gives a well-circumscribed, smooth surface of tan, white, firm-gritty material. The tumors range in size from a few millimeters up to 10 cm.

A hematoxylin and eosin stained slide show a cellular stroma with an innumerable psammoatoid calcifications. By microscopic evaluation, the tumors are composed of a variably cellular stroma make up of spindled to stellate fibroblast-like cells. Within this stroma, are numerous small, rounded, mineralized collagenous ossicles and immature osteoid. Many times the curved-shaped bone fragments have a collagenous rim around them. Ossicles may fuse to form much large mineralizations. Cementum-like psammomatous bodies (cementicles) may also be present. Osteoblastic rimming is not uncommon. Occasionally, giant cells and even mitoses are seen. It is important to get complete excision as early in the disease process as possible. Once the lesion is removed, the prognosis is excellent. However, if the lesion is incompletely excised, recurrences may be seen in up to 60% of patients. The recurrences are highest in sinus tumors [8].

The vast majority of fibro-osseous lesions are incidental findings on radiographs. They are slow-growing tumors, and are infrequently symptomatic. frontal headache is the most common complaint, followed by drainage and nasal obstruction associated with secondary sinusitis. The present study was planned to assess the symptomatic fibro-osseous lesions of the paranasal sinuses.

Methodology

The study was planned in the Department of ENT in Nalanda Medical College, Patna from Jan 2018 July 2018. Total 20 patients diagnosed with the fibro osseous lesions were enrolled in the present study. The information regarding patient demographic characteristics, location of the lesion at presentation, CT scan findings, management approach, details of surgery performed, and outcomes were collected and presented as below.

All the patients were informed consents. The aim and the objective of the present study were conveyed to them. Approval of the institutional ethical committee was taken prior to conduct of this study.

Following was the inclusion and exclusion criteria for the present study.

Inclusion criteria: Inclusion criteria were cases with symptomatic fibro osseous lesions.

Exclusion criteria: Exclusion criteria were asymptomatic cases with incidental radiological findings

Results & Discussion

Fibro-osseous lesions are benign tumours characterised by the replacement of normal bone by a fibrous cellular stroma containing various amounts of foci of mineralisation or ossification. These lesions rarely affect the sinonasal tract. They are usually divided into 3 different entities, namely osteoma, fibrous dysplasia (FD) and ossifying fibroma (OF). Osteoma is the most frequent subtype, followed by FD and OF. The precise incidence of OF, however, remains unknown. These three entities share several clinical, radiological and histological similarities, but have different behaviours in the sinonasal tract. Osteoma is a slow-growing tumour, and FD usually stops growing spontaneously after puberty [9-10].

Table 1: Demographic characteristics of patients

	Osteoma	Fibrous dysplasia (FD)	Ossifying Fibroma (OF)	Juvenile active ossifying fibroma (JAOF)
Number of patient	10	7	2	1
Gender				
Male	7	3	1	1
Female	3	4	1	0
Age (years)	7 – 40	5 -33	18 – 26	17

Table 2: Clinical characteristics

Lesion site	Osteoma No. of Cases
Sphenoid	2
Ethmoid	2
Frontal	2
Fronto-ethmoid	2
Craniofacial	1
Frontal and skull base	1
Procedure	
Endoscopic sinus surgery	8
Debulking	2
Complications	
Yes	0
No	10
Outcome	
No recurrence, improve	10
Recurrence	0

Table 3: Summary of data for patients with fibrous dysplasia/ossifying fibroma/JAOF.

	Fibrous dysplasia (FD)	Ossifying Fibroma (OF)	Juvenile active ossifying fibroma (JAOF)
Lesion site			
Sphenoid	2	0	0
Maxillary	1	0	0
Fronto-ethmoid and skull base	1	0	0
Fronto-ethmoid	2	0	0
Craniofacial	1	0	0
Ethmoid, orbit, skull base	0	1	0
Ethmoid, orbit, middle turbinate	0	1	1
Procedure			
Functional endoscopic sinus surgery (FESS)	7	1	0
FESS+lateral rhinotomy +craniotomy	0	1	0
FESS, craniotomy	0	0	1
Complications			
Yes, temporary diplopia, enophthalmos	0	1	0
No	7	1	1
Outcome			
No recurrence	7	1	0
Recurrence	0	1	1

This clinical series demonstrates the numerous ways in which fibrous dysplasia involving the skull base can present. In many respects, the series differs from previous reports on fibrous dysplasia. Determining the true incidence of fibrous dysplasia, particularly for the more prevalent monostotic form, is difficult because many patients are asymptomatic and are often diagnosed incidentally after radiographic evaluation for other reasons. Onset is typically in adolescence or late childhood, although more severe forms can arise in infancy.

Pathologically, fibrous dysplasia lesions are characterized by expansion of cortical bone with gradual replacement by fibrous tissue that is firm, rubbery, and gritty. Lesions within the skull tend to have a firmer consistency than their counterparts in the long bones of the body due to a greater amount of bony spicules [11]. Cystic lesions can often be filled with an amber fluid and can occasionally be vascular. Microscopically, the lesion is readily identifiable, with an irregular trabeculae of woven bone intermixed with a connective tissue stroma. Lesions will vary in the amount and distribution of bone and in the cellularity and vascularity of the fibrous stroma.

Fibro-osseous tumors are the most frequent tumors arising in the frontal sinus and frontal recess. Of these, the most common is the osteoma. In 1941, Wallace Teed credited Veiga with the first description of a frontal sinus osteoma in 1506, whereas Vallisnieri was credited with detailing their bony origin [12]. The frequency of frontal sinus osteomas has been known for many years as Childrey, in 1939 cited an incidence of 0.43% in 3510 skull radiographs. More recently, osteomas were found in 1% of frontal sinus radiographs in symptomatic individuals [13-14]. These bony tumors typically present in the third to fourth decade of life with a male to female ratio of 1.5:1 to 2:1. In patients of Middle Eastern or West Indian descent they may present earlier [15]. The most common presenting symptoms are headache and pain in the frontal area; however, many tumors are asymptomatic and are detected on imaging obtained for other reasons. Symptoms consistent with frontal sinusitis due to outflow obstruction are also common. With larger tumors, facial cosmetic deformity may result from anterior growth, while proptosis, diplopia, and visual changes may result from inferior extension. Posterior extension may lead to intracranial complications [16].

Polyostotic fibrous dysplasia was first described by Albright in 1937, and ossifying fibroma was distinguished from it in 1963 by Reed [17-18]. In contrast to osteomas, these lesions tend to occur in a younger population. Both fibrous dysplasia and ossifying fibroma are less frequently found in the region of the frontal recess, and they tend to be less well localized. It is for this reason that resection of a focus of fibrous dysplasia tends to require multiple attempts.

Ossifying fibroma has a tendency to recur more so than osteomas but less so than fibrous dysplasia [19]. Furthermore, pain tends to be less common whereas facial asymmetry and cosmetic deformity are more common. Of note, radiation is avoided in the treatment of fibrous dysplasia due to the risk of malignant transformation.

Ossifying fibroma (OF) is the most concerning of the fibro-osseous lesions. It is known as cemento-ossifying fibroma, psammomatoid, OF, and JAOF. OF have been more frequently observed in the third and fourth decades of life and has a higher incidence in the black population [20].

A clinical variant of OF, called juvenile-aggressive ossifying fibroma, is more frequently observed in the sinonasal tract, where it predominantly affects male subjects. It usually belongs to the cementiform type and is considered to have an aggressive biological behavior mimicking a malignant neoplasm. Even though painless facial and skull deformities are the most frequently observed signs, symptoms such as nasal obstruction, headache, epistaxis, anosmia, loosening of teeth, facial paralysis, hearing loss, trigeminal neuralgialike pain, and recurrent rhinosinusitis due to drainage impairment may develop.

Conclusion:

The data generated from the present study concludes that Diagnostic dilemma of benign fibro-osseous lesions of the paranasal sinuses and skull base can be resolved with a combination of clinical, radiological and pathological criteria. Correct diagnosis of fibroosseous tumors is crucial for adequate therapy as their treatment, prognosis, clinical aggressiveness and outcomes of individual entities vary significantly.

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