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Evaluation of fibro-osseous lesions of the paranasal sinuses

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Abstract
Fibro-osseous lesions are a diverse group of processes that are characterized by replacement of normal bone by fibrous tissue containing a newly formed mineralized product. The designation fibro-osseous lesion is not a specific diagnosis and describes only a process. Fibro-osseous lesions of the jaws include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms. The pathologic features may be very similar in lesions of diverse cause, behaviour and prognosis. Clinical, radiographic and histopathological correlation is usually most beneficial in establishing a specific diagnosis. These lesions can be classified as fibrous dysplasia, ossifying fibroma (of) and cement-osseous dysplasia. Hence based on above findings the present study was planned for Evaluation of Fibro-osseous Lesions of the Paranasal Sinuses.
The present study was planned in Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India. In the present study 15 patients diagnosed with the fibro osseous lesions were enrolled. 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 Fibrous Dysplasia is significant for the otorhinolaryngology because it may affect facial and cranial bones and may cause deformities and dysfunctions. In spite of its benign nature, the signals and symptoms resulting from the compression of noble structures of the cranial base and orbit may generate diagnostic doubts as for the possibility of a malignant lesion. The surgical treatment must take into account the disease's harmful and recurrent potential, by choosing a more conservative approach and removing as much tissue as possible to prevent mutilations and functional deficits.

Keywords: fibro-osseous lesions, osteoma, fibrous dysplasia (fd), ossifying fibroma (of), juvenile active ossifying fibroma (jaof), etc.

Introduction
Fibrous dysplasia 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. Disease occurs along a broad clinical spectrum ranging from asymptomatic, incidental lesions, to severe disabling disease. Disease can affect one bone (monostotic), multiple (polyostotic), or all bones (panostotic) 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. Fibrous dysplasia is very rare, and there is no known cure. Fibrous dysplasia is not a form of cancer.
ibrous dysplasia represents about 5% of benign bone lesions [3]; however, the true incidence is unknown, as many patients are asymptomatic. Monostotic fibrous dysplasia accounts for 75-80% of the cases.
Fibrous dysplasia is a slowly growing lesion that usually appears during periods of bone growth and is thus seen in those in early teen and adolescent years. Polyostotic fibrous dysplasia accounts for 20-25% of cases, and patients tend to present at a slightly earlier age (mean age, 8 y) [5].
Pregnancy can cause increased growth of the lesion as well as secondary changes of aneurysmal bone cyst formation. However, males and females are equally affected, although the polyostotic variant associated with McCune-Albright syndrome is seen more frequently in females [3].
Patients with small, monostotic lesions may be asymptomatic, with the osseous abnormality identified incidentally on radiologic studies obtained for unrelated reasons. However, bone pain, swelling, and tenderness are common presentations in symptomatic patients [4].
Endocrine disturbances may be the initial presentation in some patients. This section will briefly review the common affected locations of fibrous dysplasia, fibrous dysplasia deformity and fracture, and malignant transformation for this lesion.
The most common sites of skeletal involvement in monostotic fibrous dysplasia are the ribs, proximal femur, and craniofacial bones, typically the posterior maxilla [4, 5]. The lesion may involve only a small segment of bone or it may occupy its entire length.
In polyostotic fibrous dysplasia, the spectrum of involvement varies from 2 bones to more than 75% of the skeleton. Polyostotic fibrous dysplasia is most commonly found in the femur, tibia, pelvis, and foot. Other sites less commonly affected include the ribs, skull, and bones of the upper extremity. Uncommonly affected bones include the lumbar spine, clavicle, and the cervical spine.
The most common physical deformities are leg-length

discrepancy, facial asymmetry due to hemicranial involvement, and rib deformities.

Fracture is the most common complication in fibrous dysplasia [6]. It is seen in more than half of the patients with the polyostotic form of the disease. Deformities in weight-bearing bones can occur. Almost 75% of patients with polyostotic fibrous dysplasia are symptomatic, with pain, deformity, or pathologic fractures [3].

Malignant transformation of fibrous dysplasia occurs very infrequently, with reported prevalences ranging from 0.4% to 4% [6]. Previous irradiation has been documented in more than half of the cases with malignant transformation.

The most common malignant tumors are osteosarcoma, fibrosarcoma, and chondrosarcoma, and the majority of patients are older than thirty years when the sarcoma is diagnosed. The craniofacial region is the most common site of involvement, followed by the femur, tibia, and pelvis. The rate of malignant transformation is higher for polyostotic lesions than for monostotic lesions.

On plain films, fibrous dysplasia is an intramedullary, expansile, and well-defined lesion in the diaphysis or metaphysis. The lesions can vary from completely radiolucent to completely sclerotic; however, most lesions have a characteristic hazy ground-glass appearance (see the following images) [3]. The degree of haziness shown radiographically by a given lesion correlates directly with its underlying histopathology. More radiolucent lesions are composed of predominantly fibrous elements, whereas more radiopaque lesions contain a greater proportion of woven bone.

Fibrous dysplasia is a noninherited developmental anomaly of bone in which normal bone marrow is replaced by fibro-osseous tissue [1, 2]. This condition was first described in 1942 by Lichtenstein and Jaffe [3]; hence, fibrous dysplasia is sometimes referred to as Lichtenstein-Jaffe disease. The disease process may be localized to a single bone (monostotic fibrous dysplasia) or multiple bones (polyostotic fibrous dysplasia) [4].

Polyostotic fibrous dysplasia can occur as a part of McCune-Albright syndrome (unilateral polyostotic fibrous dysplasia, ipsilateral café-au-lait spots on the skin, and endocrine disturbances such as precocious puberty) or Mazabraud syndrome (polyostotic fibrous dysplasia and soft-tissue myxomas). Fibrous dysplasia has also been reported in association with other endocrine dysfunctions, [4] such as hyperthyroidism, hyperparathyroidism, acromegaly, diabetes mellitus, and Cushing syndrome.

At the patient's initial presentation, scintigraphy may be used to demonstrate the extent of disease. Active fibrous dysplasia lesions in younger patients have greatly increased isotope uptake; the uptake becomes less intense as the lesions mature.

The extent of the lesion is best demonstrated on computed tomography (CT) scans. This imaging modality is helpful in distinguishing fibrous dysplasia from other lesions in the differential diagnosis.

Magnetic resonance imaging (MRI) is a sensitive means of establishing the lesion's shape and content [7]. Because fibrous dysplasia is composed mainly of fibrous tissue and bone, T1-weighted images have a low-intensity signal [6]. T2-weighted images have a higher intensity signal that is not as bright as the signal of malignant tissue, fat, or fluid.

Incidentally discovered, asymptomatic, radiographically characteristic fibrous dysplasia lesions do not require further assessment and require only clinical observation [8]. Follow-up radiographs every 6 months to look for progression has been recommended. In newly identified cases, a bone scan is needed to exclude a diagnosis of polyostotic disease. When polyostotic disease is found, referral to an endocrinologist for early detection of possible systemic abnormalities is warranted.

Bisphosphonates, primarily intravenous pamidronate, have been utilized to decrease bone pain in symptomatic patients with polyostotic disease [8].

Open biopsy may be indicated to confirm the diagnosis of fibrous dysplasia when there is a nonclassic presentation. Surgical procedures are required for correction of deformities, prevention of pathologic fractures, or eradication of symptomatic lesions [9].

Treatment of malignant transformation is based on the subtype of sarcoma, but the prognosis tends to be worse for patients with malignant transformation than it is for those with a similar primary sarcoma.

The macroscopic appearance of fibrous dysplasia is that of a centrally located, tan to gray white, gritty-feeling lesion (see the image below). Areas of cyst formation or hemorrhage may be identified. The lesion can be easily peeled away from the encircling shell of reactive bone by blunt dissection. Occasionally, small islands of cartilage that undergo calcification and enchondral ossification may be observed in the lesional tissue. This phenomenon is most frequently encountered in the proximal femur and is sometimes referred to as fibrocartilaginous dysplasia.

Histologically, fibrous dysplasia is composed of fibrous tissue with randomly oriented bony trabeculae that are thought to be formed by osseous metaplasia of the fibrous stroma [6].

The fibrous stroma is usually of low cellularity and contains variable amounts of myxoid material to dense collagenous matrix [10, 11]. The fibroblasts usually have plump, ovoid nuclei, although elongated, narrow nuclei may also be seen. A storiform pattern of the fibroblasts may also be identified [12].

The osseous trabeculae are composed of immature woven bone and are typically not lined by osteoblasts, although focal osteoblastic rimming may be seen. Osteoclasts are frequently observed, especially on the concave side of the trabeculae. The outline of the trabeculae varies from solid, round islands to short, irregular, curvilinear or serpiginous shapes, giving the characteristic "Chinese character" or "alphabet soup" appearance [12]. The ratio of fibrous tissue to bone ranges from fields that are totally fibrous to those filled with dysplastic trabeculae [3, 6].

Multiple, delicate capillaries are found throughout the lesion and, when injured, incite a giant-cell reactive process. A cartilagenous component, composed of lobules of mature hyaline cartilage (fibrocartilaginous dysplasia), may be encountered, most often in the proximal femur. Hemorrhage and cystic change may be occasionally found. Secondary aneurysmal bone cyst formation can also occur.

Fibro-osseous lesions are a diverse group of processes that are characterized by replacement of normal bone by fibrous tissue containing a newly formed mineralized product. The designation fibro-osseous lesion is not a specific diagnosis and describes only a process. Fibro-osseous lesions of the jaws include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms. The pathologic features may be very similar in lesions of diverse

cause, behaviour and prognosis. Clinical, radiographic and histopathological correlation is usually most beneficial in establishing a specific diagnosis. These lesions can be classified as fibrous dysplasia, ossifying fibroma (OF) and cement-osseous dysplasia. Hence based on above findings the present study was planned for Evaluation of Fibro-osseous Lesions of the Paranasal Sinuses.

Methodology

The present study was planned in Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India. In the present study 15 patients diagnosed with the fibro osseous lesions were enrolled. 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 of the jaws represent a diverse group of entities which include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms [13]. Benign fibro-osseous lesions of the head and neck region are uncommon and represent a wide range of tumors sharing some histopathological features; which comprise fibrous dysplasia, ossifying fibroma (of), and cement-osseous dysplasia. OF can be further divided into conventional and juvenile forms (juvenile ossifying fibroma (JOF)) [14].

Benjamins in 1938 first reported PsJOF as an "osteoid fibroma with atypical ossification of the frontal sinus", [15] Golgi in 1949 called it as "Psammomatoid Ossifying Fibroma", and Johnson in 1952 coined the term "Juvenile Active Ossifying Fibroma". According to WHO classification of odontogenic tumors 2005, it was named as "Juvenile Psammomatoid Ossifying Fibroma". The term juvenile psammomatoid ossifying fibroma is now used to designate the neoplasm of the craniofacial skeleton of young age with well-defined clinicopathological features [16].

The pathogenesis of these jaw lesions are related to the abnormal development of basal generative mechanism that is essential for root formation [15].

Table 1: Demographic characteristics of patients

	Osteoma	Fibrous dysplasia (FD)	Ossifying Fibroma (OF)	Juvenile active ossifying fibroma (JAOF)
Number of patient	8	5	1	1
Gender				
Male	6	3	1	1
Female	2	2	0	0
Age (years)	5 – 39	6 - 43	7 – 36	5 - 41

Table 2: Clinical characteristics

Lesion site	Osteoma No. of Cases
Sphenoid	2
Ethmoid	2
Frontal	1
Fronto-ethmoid	1
Craniofacial	1
Frontal and skull base	1
Procedure	
Endoscopic sinus surgery	6
Debulking	2
Complications	
Yes	0
No	8
Outcome	
No recurrence, improve	8
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)
Total Cases	5	1	**1**
Lesion site			
Sphenoid	2	0	0
Maxillary	1	0	0
Fronto-ethmoid and skull base	1	0	0
Fronto-ethmoid	1	0	0
Craniofacial	0	0	0
Ethmoid, orbit, skull base	0	1	0
Ethmoid, orbit, middle turbinate	0	0	1
Procedure			
Functional endoscopic sinus surgery (FESS)	5	1	0
FESS+lateral rhinotomy+craniotomy	0	0	0
FESS, craniotomy	0	0	1
Complications			
Yes, temporary diplopia, enophthalmos	0	0	0
No	5	1	1
Outcome			
No recurrence	5	0	0
Recurrence	0	1	1

Fibrous dysplasia and ossifying fibroma are locally destructive and deforming benign lesions that can occur anywhere in the craniofacial skeleton, including the paranasal sinuses. They are encompassed within the

'umbrella' term 'fibro-osseous lesion' (Commins DJ, 1998 & Dornhoffer J, 1995) [17-18].

Fu and Perzin (1974) noted 9 cases of fibrous dysplasia among 256 cases of non-epithelial tumors involving the nasal cavity, paranasal sinuses and nasopharynx. 6 patients were females and 3 were males with an average age of 22 years [19].

Fibrous dysplasia of ethmoid was found in only two (2) cases. Von Rampaey *et al* (1994) [20] described only 3 cases of fibrous dysplasia and ossifying fibroma of paranasal sinuses. Dornhoffer & Schwager (1995) have found only 2 cases of fibrous dysplasia and ossifying fibroma of paranasal sinuses. Conservative surgery is recommended for fibrous dysplasia of craniofacial bones (Pinsolle V *et al*, 1998) [21].

Ossifying fibroma is a type of fibro-osseous lesion of the jaws arising from the undifferentiated cells of the periodontal ligament affecting the tooth-bearing areas especially premolars and molars. It occurs mostly in adults. Its relatively more aggressive variant predominant in children below 15 years of age has been termed as juvenile ossifying fibroma (JOF). Initial clinical manifestation of JOF is in the form of a swelling of the maxilla which is usually asymptomatic. When the orbital bone and the paranasal sinuses are involved, signs such as pain along with exophthalmos, bulbar displacement and nasal obstruction appear [22]. On radiographs, the lesions appear as circumscribed radiolucencies that in some cases contain central radiopacities giving a "groundglass" or "honey-comb" appearance [23].

According to the WHO classification of odontogenic tumors 2005, JOF is further subdivided into juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF). The trabecular type is seen more commonly in children whereas the psammomatoid type has been reported in relatively adult age groups as well. The basic difference between the two types lies in the pattern of calcification of the masses of osteoid dispersed in fibrous stroma. JPOF presents with concentric calcifications imparting a psammoma body-like appearance to the spherical masses. However, JTOF shows calcification progressing into the osteoid strands, producing woven bone trabeculae that are rimmed with osteoblasts and incorporated osteocytes [24]. Differential diagnosis can be established with cemento-ossifying fibroma (clinically predominant in adults and predilection for mandible, histological variations), osteo-fibrous dysplasia and fibrous dysplasia (radiographically diffuse margins as compared to clearly defined cortical margin in JOF) [25].

There have been reports of medical management of FDs with bisphosphonates; however, the results have not been promising [26]. Some authors claim improvement of pain and inflammatory symptoms, stabilization and reduction of bone destruction, increase of the osseous density, recalcification of osteolytic lesion in 50% of the patients, improvement of the radiologic aspects and osseous metabolism.

Clinical findings of increasing pain and an enlarging soft tissue mass suggest malignant change. Rarely, a sarcoma may develop in patients with FD. Osteosarcoma is the most common type, which most often affects the craniofacial bones. Contrary to popular belief, this development has been noted in tissue, which has not received radiation as a part of the treatment in FD [27-28].

Fibroosseous lesions represent a spectrum of diseases, which could sometimes result in severe disfigurement, as seen in some of the above cases. An insight into the genetic basis of this disease process and an understanding of the clinical and radiological features is essential not only for the diagnosis but also in making a sound decision for the control of this disease in its various stages of progression.

Conclusion

The Fibrous Dysplasia is significant for the otorhinolaryngology because it may affect facial and cranial bones and may cause deformities and dysfunctions. In spite of its benign nature, the signals and symptoms resulting from the compression of noble structures of the cranial base and orbit may generate diagnostic doubts as for the possibility of a malignant lesion. The surgical treatment must take into account the disease's harmful and recurrent potential, by choosing a more conservative approach and removing as much tissue as possible to prevent mutilations and functional deficits.

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