Asymptomatic facial disfigurement – case reports of monostotic and polyostotic fibrous dysplasia

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ABSTRACT

Fibrous Dysplasia is a developmental dysplastic disorder of bone in which the normal bone matrix is replaced by fibroblastic proliferation. Von Rechlinghausen was the first author who described it in 1891 and Lichtenstein was the person who introduced the term fibrous dysplasia. It can be monostotic (70%) or polyostotic (30%). The craniofacial localization occurs in 10%-25% of cases in monostotic forms and in 50% of cases in polyostotic forms. Almost all patients with extensive polyostotic forms of the disease have skull involvement, while most patients with craniofacial bone involvement have the monostotic form of the disease. Fibrous dysplasia essentially affects children and young adults, with no sex preference. Diagnosis is not always straightforward because the functional symptomology is often absent or not specific. Only symptomatic and/or gross forms are considered for treatment.

We report two cases of fibrous dysplasia, one monostotic form involving only the maxilla and other polyostotic involving bilateral facial and skull bones.

Keywords: Craniofacial, Fibrous dysplasia, Monostotic form, Polyostotic form

INTRODUCTION

Fibrous dysplasia (FD) is a benign intramedullary fibro-osseous lesion. FD is a bone developmental anomaly characterized by replacement of normal bone and marrow bone by fibrous tissue. It is a benign intramedullary fibro-osseous lesion originally described by Lichtenstein and Jaffe in 1938. The true incidence and prevalence of fibrous dysplasia is difficult to estimate accurately, but the lesions are not rare; they are reported to represent approximately 5% to 7% of benign bone tumors. Male to female ratio is equal. It is most frequently found in metaphysodiaphyseal region of long bones i.e. in rib (28%), followed by femur (23%), tibia and craniofacial bone (10-25%). Sarcomatous transformation is rare. Some people with fibrous dysplasia have only one bone involved (monostotic), whereas other people have more than one bone involved (polyostotic). The disease may occur alone, or as part of a condition known as the McCune-Albright syndrome. McCune-Albright syndrome is characterized by fibrous dysplasia and other symptoms such as patches of pigmented skin (light brown or "café-au-lait" spots) and endocrine problems such as precocious puberty, hyperthyroidism, gigantism or acromegaly, Cushing's syndrome, and other rare conditions. 50%-100% of patient with polyostotic disease & 10% patient with monostotic disease have craniofacial involvement.[1]

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CASE DESCRIPTION AND RESULTS

Case 1: A 25 year old female reported to our department with a complaint of swelling in the posterior palatal region on the left side since 5 years.

Over the years, Swelling gradually increased in size to attain the present status. 5 months back it became symptomatic with a gradual onset of pain, which was intermittent in nature, moderate in intensity, dull and aggravated on pressure application. Patient also gave the history of nasal obstruction since 4 months and discharge from left eye since 2 months.

Extra oral examination revealed single, diffuse swelling of approximately 4×3cm in size, on the left side of face. The swelling was smooth surfaced, extending superiorly to the left infra-orbital margin and inferiorly 4cm above the angle of the mouth. Medially, it extended up to the midline of nasal bridge and laterally up to the zygomatic prominence. On palpation no local rise in temperature, non-tender and hard in consistency.

Intraoral examination revealed a hard mass in the left maxillary buccal sulcus extending from the left lateral incisor to the third molar of same side causing buccal cortical bone expansion alone (Fig. 1). Deciduous 1st& 2nd molar are retained with slight mobility. Displacement of maxillary canine buccally was seen. Overlying mucosa appeared normal and non-tender. Clinical diagnosis of fibrous dysplasia was given.

Intraoral periapical view showed a completely radiopaque lesion involving the posterior maxilla, which was ill defined, obscuring lamina dura of the adjacent teeth (Fig. 2). Prominent buccal cortical expansion was apparent on maxillary occlusal view and Computed tomography scan slices. The outer cortical plate was not intact. Computed tomography scan showed Homogenous hyperdense pattern filling the left maxillary sinus completely (Fig. 3). Lesion also infiltrated into the nasal cavity compressing the inferior nasal conchae (Fig. 4; 5). Radiological features were in concurrence with the clinical diagnosis. Histopathological examination confirmed the fibro-osseous lesion (Fig. 6)

Case 2: A 17 year old female patient reported to our department with a complaint of painless, gradually progressive swelling in the right side of the face since 5 years. It was not associated with nasal obstruction, or discharge from eye or nose.

Gross facial asymmetry was observed due to a single, diffuse swelling of approximately 4x3cm in size, in the middle region of the face on the right side. It extended supero-inferiorly from 1 cm below the infra-orbital margin to the angle of the mouth and antero-posteriorly from right lateral border of the nose, obliterating the nasolabial fold to 3 cm short off tragus. On palpation the swelling was bony hard, non-tender without local rise in temperature.

On intra oral examination, obliteration of both right and left upper buccal vestibule was evident due to a diffuse, smooth surface swelling of approximately 5x2 cm and 3×2 cm in size respectively. Lesion on the right side showed increased bucco-palatal dimension from canine region to maxillary tuberosity with the maximum dimension at the molar region, suggestive of buccal cortical bone expansion. Whereas, lesion on left showed similar changes at premolar – molar region. On palpation it was non-tender and bony hard in consistency. Clinical diagnosis of fibrous dysplasia was given.

Blood and Biochemical investigation showed Serum Calcium (9.3 mg/dl) and phosphorus (4.1 mg/dl) within normal range whereas alkaline phosphatase (240 IU/L) turned out to be raised.

Gross radio-opacity in maxillary bone on both sides was revealed on OPG and Occlusal view. Buccal Cortical bone expansion was also confirmed with occlusal view. The Computed tomography scan of craniofacial region showed bilateral hyper dense expansive mass (Fig. 7) involving multiple facial bones viz nasal, lacrimal, maxillary, zygomatic and inferior nasal conchae (Fig. 8). Among cranial bones; frontal, ethamoid, sphenoid, squamous part of occipital and temporal were involved (Fig. 9). The clinical diagnosis of fibrous dysplasia was confirmed.

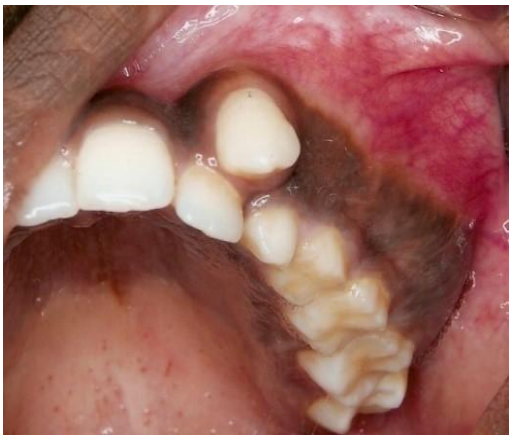

Fig. 1: Intraoral view reveals a hard mass in the left maxillary buccal sulcus

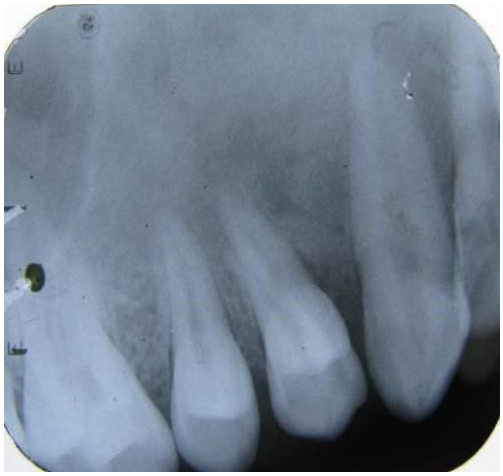

Fig. 2: Intraoral periapical view

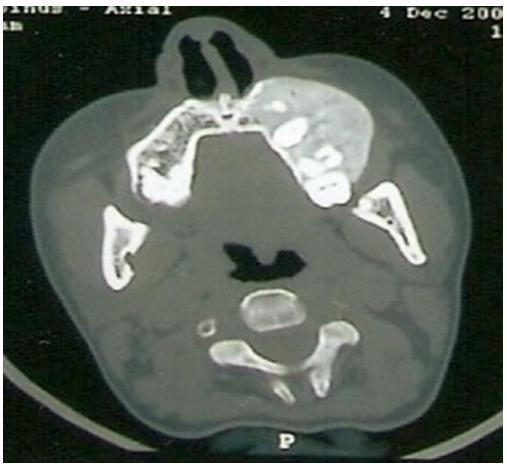

Fig. 3: Computed tomography scan showing left maxillary sinus

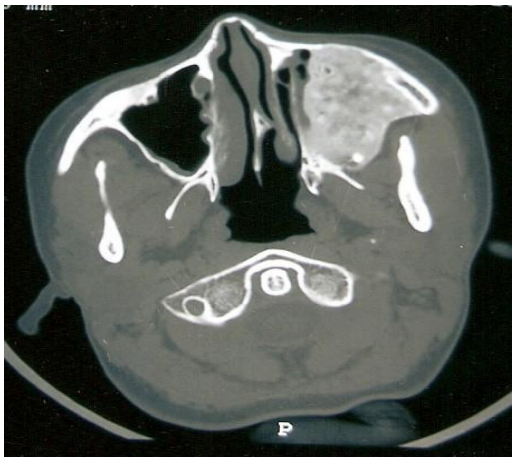

Fig. 4: Computed tomography scan showing inferior nasal conchae

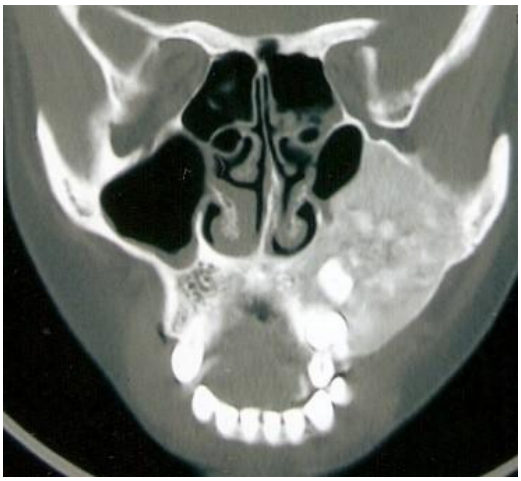

Fig. 5: Computed tomography scan showing inferior nasal conchae

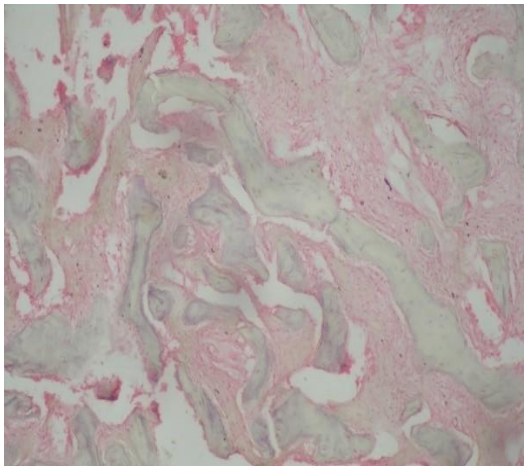

Fig. 6: Histopathological view

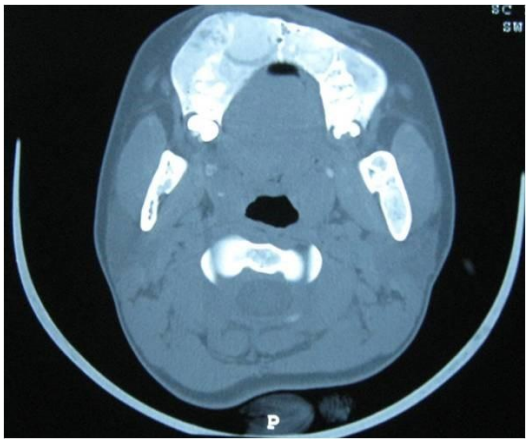

Fig. 7: Computed tomography scan of craniofacial region

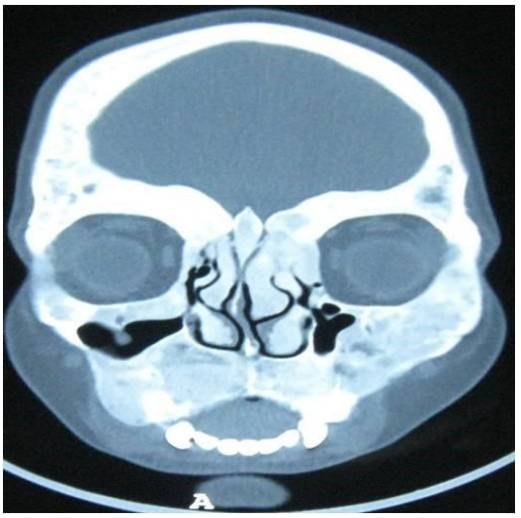

Fig. 8: Computed tomography scan showing nasal, lacrimal, maxillary, zygomatic and inferior nasal conchae

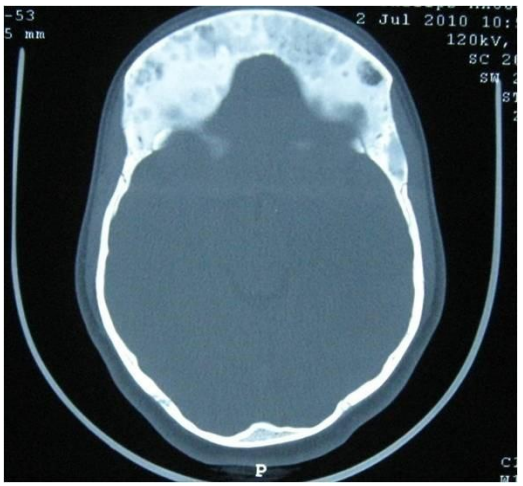

Fig. 9: Computed tomography scan showing frontal, ethamoid, sphenoid, squamous part of occipital and temporal bones

DISCUSSION

The etiology of fibrous dysplasia has been linked with a mutation in the Gsα gene that occurs after fertilization in somatic cells and is located at chromosome 20q13.2-13.3. Fibrous dysplasia is postulated to occur as a result of a developmental failure in the remodeling of primitive bone to mature lamellar bone and a failure of the bone to realign in response to mechanical stress. Failure of maturation leaves a mass of immature isolated trabeculae enmeshed in dysplastic fibrous tissue that are turning over constantly but never completing the remodeling process.[2]

The clinical presentation depends on the site, duration, extent and nature of the lesion, which can affect any bone in the body. It ranges from a mild local swelling with little or no pain to a gross deformity with complications such as proptosis, visual disturbance and sensorineural hearing loss. Maxilla is more commonly involved than mandible. When maxilla is affected it may involve zygomatic & sphenoid bone. The pattern of which bones are involved is established very early in life and does not change with age.[3]

The plain film radiological features of FD are non-specific and vary widely. The normal bone is replaced by tissue that is more radiolucent, with a grayish "ground-glass" pattern that is similar to the density of cancellous bone but is homogeneous, with no visible trabecular pattern. Fries, has described three radiological patterns in craniofacial fibrous dysplasia. The first is pagetoid with bone expansion and alternate areas of radiopaque and radiolucency. It occurs in more than half of the patients, most of who are older than 30 years of age and have had symptoms for an average of 15 years. The second pattern is sclerotic, with bone expansion and a homogenous radio density (ground glass appearance). The third type is cyst-like, usually a round or oval lesion with a sclerotic border, which occurred in younger individuals. The incidence of pagetoidis type 56%, sclerotic type 23%, and the radiolucent type 21%. The most effective method of monitoring growth and estimating extent of disease seems to be computerized tomography. Petrikowski et al. suggested that the loss of lamina dura could be used as an ancillary diagnostic feature for Fibrous dysplasia.[4]

The decalcified, H&E stained sections show densely packed irregular shaped trabeculae of lamellar bone, showing lacunae and osteocytes, collagen matrix stroma with fibroblasts in an entangled standard with osseous trabeculate similar to the "Chinese writing", with no osteoblastic rimming. We described above includes a symptomatic and asymptomatic cases. The differential diagnosis includes Odontogenic cysts, Central ossifying fibroma, Aneurysmal bone cyst, Sclerosing Osteomyelitis, paget's disease.[5,6]

Although there was no compelling indication to seek a biopsy, any sudden change in the clinical presentation or behavior of the lesion might warrant further investigation. Malignant transformation of fibrous dysplasia ranges from 0.4% to 4%. The recurrence rate of fibrous dysplasia falls within the range of 0 to 13.3%.[6] The main aim of the treatment is correction of the functionality in combination with aesthetic effects. The conservative therapeutical approach with limited reduction in the size of these lesions is enough to manage the symptoms. The radiotherapy increased malignant transformations more than 400 times.[7]

CONCLUSION

Fibrous Dysplasia is a benign disease that has the potential to cause significant cosmetic and functional disturbance. Much progress has been made over the past decade, for example the identification of the genetic mutation linked to the etiology of the disease. With proper understanding, diagnosis and management, good outcomes can often be achieved. Awareness of the myriad of radiographic appearances of fibrous dysplasia is essential and will be beneficial in the accurate diagnosis and proper treatment planning, even without invasive diagnostic procedures.

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