International Journal of Advances in Medicine
Jagdale A et al. Int J Adv Med. 2016 Nov;3(4):1068-1073
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: http://dx.doi.org/10.18203/2349-3933.ijam20163751

Case Report

Fibrous dysplasia: a case series of five cases

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Received: 06 July 2016
Revised: 17 July 2016
Accepted: 06 August 2016

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ABSTRACT

Fibrous dysplasia is development anomaly in which normal bone marrow is replaced by fibro-osseous tissue which characterized by deformities of the bone, fractures, nerve compression and bone pain. It is most commonly seen in young adult. In this study we report five case of fibrous dysplasia occurring at various locations with radiological interpretation.

Keywords: Bones, Cranium, Fibrous dysplasia, Mono-stotic, Poly-ostotic

INTRODUCTION

Fibrous dysplasia is a developmental anomaly in which normal bone marrow is replaced by fibro-osseous tissue.[1,2] It is a skeletal developmental disorder of bone forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation.[3,4]

Reed defined fibrous dysplasia as an arrest of bone maturation.[3] It is a rare disorder characterized by deformities of bone, fractures, nerve compressions and bone pain.[5] It can affect a single bone, or even a small segment or the entire skeleton diffusely.[1,6]

Fibrous dysplasia is a benign skeletal disorder typically seen in young adults.[1,7] it is not a true neoplasm.[1]

Fibrous dysplasia as a separate entity was first described by Lichenstein and Jaffe and hence is also called as Lichtenstein Jaffe disease.[1-3,8,9]

It is associated with many endocrinological diseases, most common being Albright syndrome.[7,8] There is no familial or hereditary or congenital basis to the disease.[4,9]

Clinical features

Bone pain is the most common feature.[5]It is mostly a disease of young age, mostly seen in first and second decade.[1,4,8]

Other features are

Cutaneous lesions in the form of café-au-lait spots. The borders of these lesions are typically irregular or serrated (Coast of Maine).[1] Facial asymmetry can be seen due to hemi-cranial involvement.[1] Pregnancy can exacerbate fibrous dysplasia and also cause aneurysmal bone cyst formation.[1] Sinusitis.

Patho-physiology

It is thought to occur as a result of abnormal activity of mesenchymal cells.[4,9] It occurs as a result of mutation in the gene that encodes the sub-unit of a stimulatory G protein (GS alpha) located on chromosome 20.[2,3,8,10] There is substitution of cysteine by arginine.

Lesions of fibrous dysplasia are composed of fibrous tissue containing bone trabeculae.[1,10] Fibrous stroma is a

myxofibrous tissue of low vascularity while the bony trabeculae are composed of woven bone. [1] The outline of the bone trabeculae has been likened to the Chinese characters or alphabet soup.[1,2]

There is no osteoblastic activity, while osteoclasts are typically seen, especially on the concave side of trabeculae.[1,10] Lesions of the fibrous dysplasia are characterized by expansion of cortical bone with gradual replacement by fibrous tissue that is firm, rubbery and gritty.[8]

CASE REPORT

CT examination of the brain, face, PNS and lower limbs was performed on Siemens Somatom Emo 6 machine with 6 mm and 2 mm sections. In two patients who had complaints in the limbs, MRI was also done. MRI was performed on 1.5T magnet MR system (Siemens magnetom Essenza). Five patients presented to the department with following findings –

CT findings

Case 1

A 35 years old female came with complaints of headache, blurring of vision and focal increase in size of the left zygoma. CT scan was performed and a 2.2 x 1.5 cm sized rounded lesion arising from the left zygoma showing ground glass matrix, suggesting a diagnosis of fibrous dyplasia.

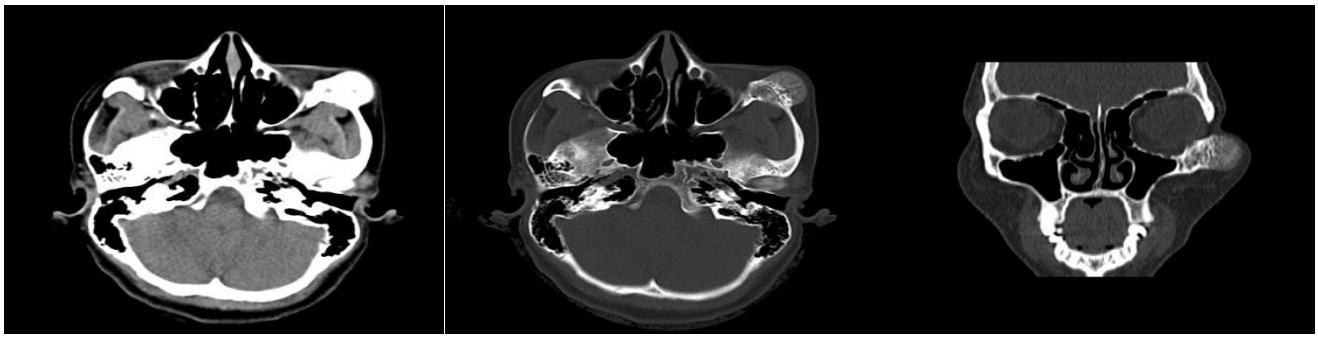

Figure 1: Rounded ground glass density arising from the left zygoma.

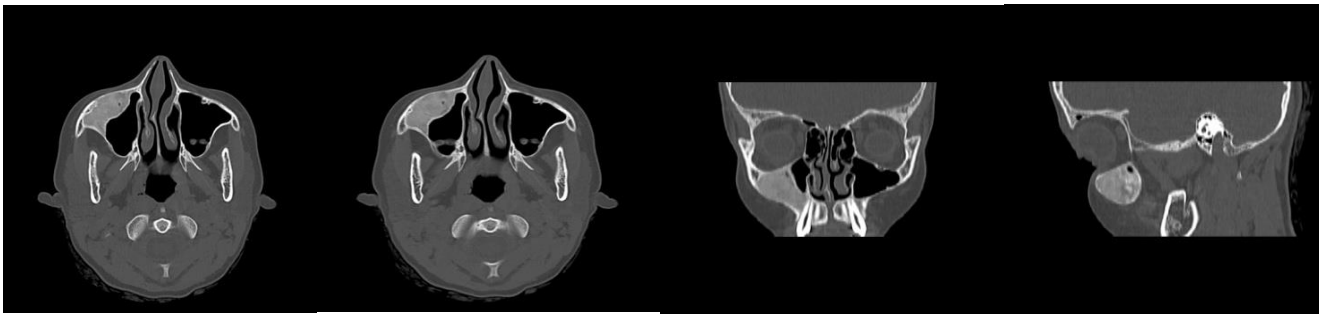

Figure 2: Diffuse osseous hyper density seen along the anterior and lateral wall of maxillary sinus.

Case 2

A 12 year old female came with complaints of sinusitis, headache, persistent nasal congestion. CT scan was performed and diffuse osseous hyperdensity (ground glass opacity) seen mainly along the anterior and part of lateral wall of right maxillary sinus. The right maxillary sinus cavity was partially encroached upon. Bony walls were intact. Features were suggestive of fibrous dysplasia.

Case 3

A 30 years old female came with complaints of gradual increase in size of the left half of face since past two to three years which was associated with gradual loss of hearing along with dysphagia. The patient also complained of motor symptoms in the form of hemiplegia on the right side. Other symptoms were forgetfulness.

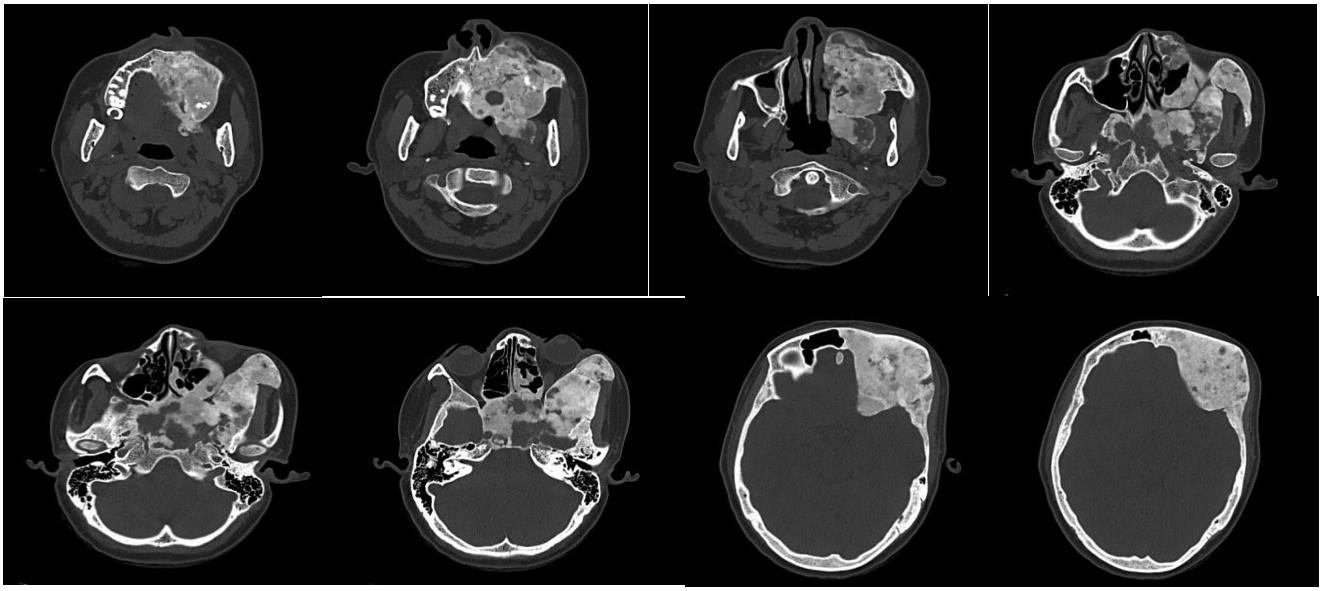

Figure 3: Ground glass density extending to the cranio-facial region s/o cranio-facial fibrous dysplasia.

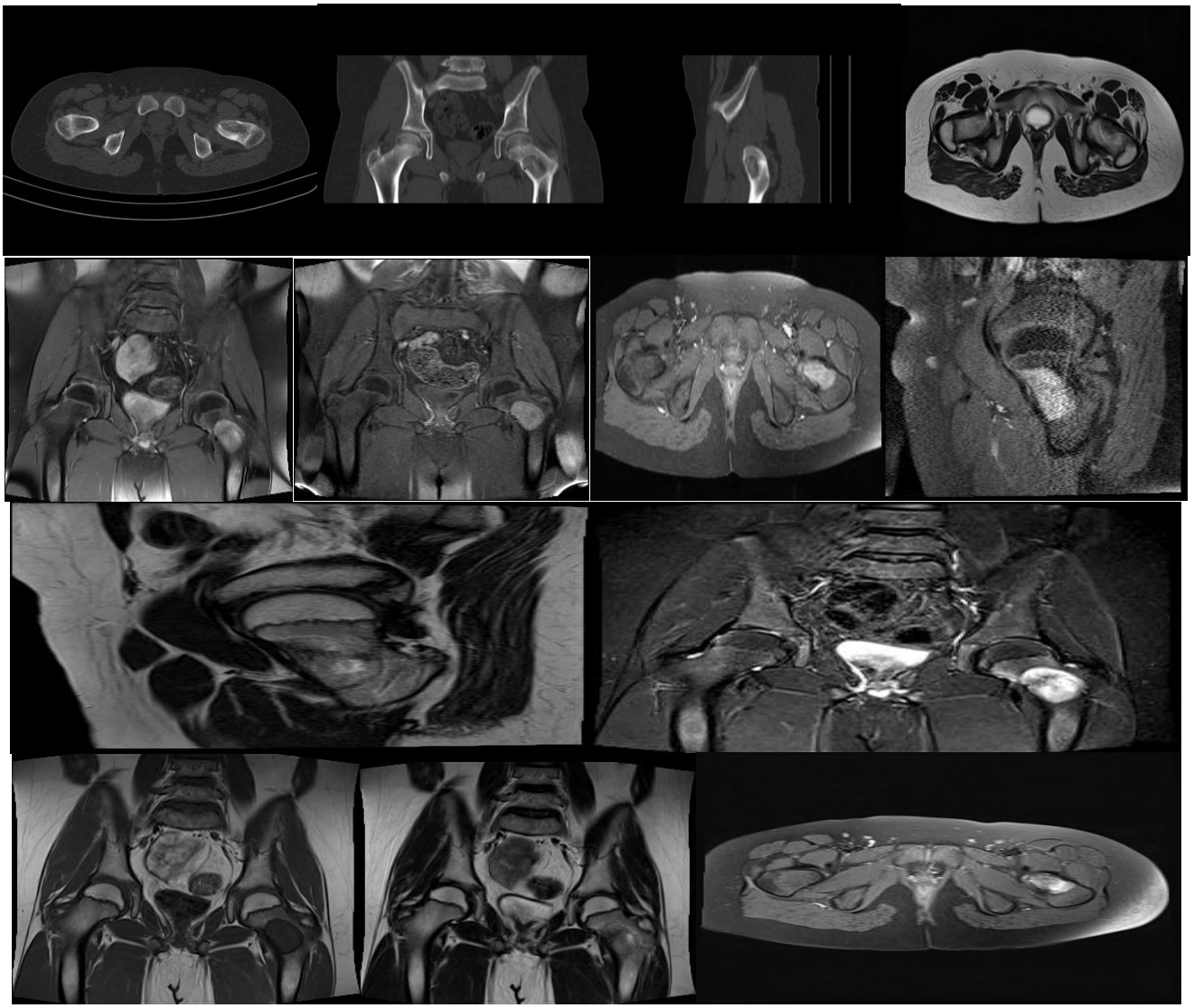

Figure 4: CT and MRI images of fibrous dysplasia arising from the neck of the left femur.

CT scan was performed on this patient and it showed ground glass opacity with multiple cystic spaces with extending from the left half of mandible to left zygoma, roof of orbit on the left side, left superior and inferior nasal turbinates, left crista galli, left sided maxillary sinus entirely, basi-sphenoid, left sided frontal bone, left side squamous and petrous temporal bone. Features were suggestive of cranio-facial fibrous dysplasia.

Case 4

12 years old male came to the department with complaints of H/O fall two years back with pain and swelling in the hip on left side since then. The patient also complained of gradually increasing limping on walking.

CT and MRI examination was performed on this patient. CT showed ground glass opacity in the neck of left femur.

MRI examination showed well demarcated ovoid lesion with narrow zone of transition in the left femoral neck anteriorly with subtle cortical irregularity in anterior cortical margin.

This lesion was hypointense on T1 and heterogeneously hyperintense on T2. Post contrast study showed mild to moderate enhancement in the lesion.

Case 5

A 25 years old male patient came to the department with complaints of gradually increasing pain in both lower limbs, parasthesias in both lower limbs and difficulty in walking.

CT and MRI examination was performed on this patient. CT showed ground glass opacity in the femoral and tibial shaft.

MRI showed expansile T2 and STIR iso to hyperintense and T1 hypointense mild heterogeneous lesions involving postero-lateral aspect of femur.

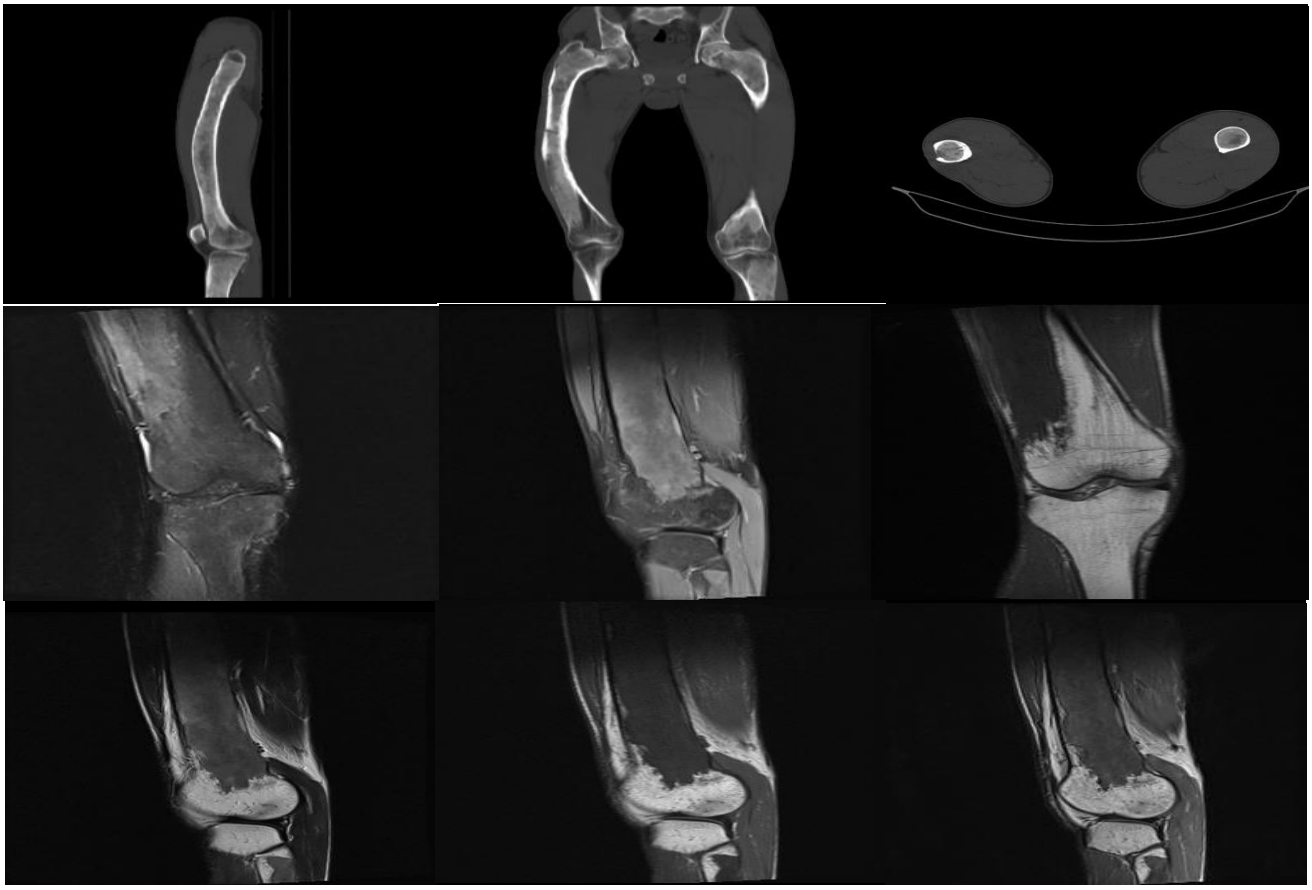

Figure No 5: CT and MRI images of fibrous dysplasia arising from the femoral and tibial shaft mostly involving postero-lateral aspect of femur.

RESULTS

Patient one had involvement of left zygoma. Patient two had involvement anterior and part of lateral wall of right maxillary sinus

Patient three had involvement of left half of mandible to left zygoma, roof of orbit on the left side, left superior and inferior nasal turbinates, left crista galli, left sided maxillary sinus entirely, basi-sphenoid, left sided frontal bone, left side squamous and petrous temporal bone. Patient four showed involvement of neck of left femur. Patient five had involvement of femoral and tibial shaft.

In this study, out of the five patients, four patients presented with mono-ostotic form and only one patient had poly-ostotic form. The involvements of the facial bones were more commonly observed followed by femoral bone.

DISCUSSION

Fibrous dysplasia is a developmental anomaly in which normal bone marrow is replaced by fibro-osseous tissue.[1,2] It is a skeletal developmental disorder of bone forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation.[3,4]

It constitutes 2 % of all bone tumors, and 7 % of all benign bony tumors.[2,9] The most common locations are ribs, femur, tibia, fibula and facial bones.[1,4,9] Skull and facial bones are affected in 10- 25 % of monostotic form and 50% of polyostotic form.[3,6] Amongst the facial bones, maxilla is most commonly involved.[9] Other sites are temporal bone and sphenoid bone.[8]

It is of two types

Monostotic

It is the most common form (70%), [3,8] and it affects single bone.[1,2,4,7]

Polyostotic

Multiple bones are involved. Female predominance is noted in polyostotic form.[3]

Polyostotic form can be associated with soft tissue myxoma.[1] The soft tissue myxoma is typically intra-muscular and almost invariably multiple.

Polyostotic form has three sub types-[2]

- Cranio-facial type - only the cranio-facial complex is involved
- Lichtenstein Jaffe type - In addition to cranio-facial complex involvement, there are also cutaneous lesions in the form of café-au-lait spots.
- Albright syndrome - It is a triad of - Polyostotic fibrous dysplasia (typically unilateral).[1]

Cutaneous café-au-lait spots (Ipsilateral to bone lesions).

Precocious puberty in girls. Complete triad is very rarely seen.[1] It is the most severe form and is more common in females.[8] Monostotic form is not believed to be a precursor to the polyostotic form.[8]

CHEZRUBISM

It is a special form of fibrous dysplasia that is autosomal dominant disorder with symmetric involvement of both maxilla and mandible.[1,4] It is more severe in males and usually regresses after adolescence.[1]

Radiological features

CT

It is the investigation of choice for diagnosis and follow up.[7,8] It describes the extent of skeletal involvement.[1]

Three main features are seen in CT

- Expanded bone with ground glass pattern[2,6]
- Homogenously dense pattern[6]
- Cystic variety.[6]

Three radiographic standards in the cranial fibrous dysplasia and facial bones have been described [2,8]

- Pategoid - alternate radiodense and radiotransparent areas
- Sclerotic - homogenously dense
- Myxoid.

Areas of low enhancement and cyst formation aids in differentiating the lesion from malignancy.[7]

MRI

Most of the lesions appear hypointense on T1WI and variable intensity on T2WI.[1,6] The variable intensity on T2WI resembles soft tissue tumor.[6] Localized fibrous dysplasia on MRI mimics a tumor because fibrous tissue enhances brilliantly after contrast administration.[6] High intensity on T2WI corresponds to non-mineralised areas and region of cystic changes seen on CT.[6]

X RAY

Lesion appears to be eccentric and medullary.[1]

The normal architecture of bone is altered and remodelled by fibrous dysplastic lesion, which is characterized by delicate woven bone spicules (ground glass appearance).[1]

Other features on X RAY[1]

- Endosteal scallping of the cortex
- Sclerotic reactive bone (rind)
- No clear demarcation with the surrounding bone.

In calvarial fibrous dysplasia

- Increased density at the base of skull
- Obliteration of sinuses
- Hemi-cranial involvement
- In femoral fibrous dysplasia.[1]

Shepherd crook deformity

Marked varus deformity. This occurs because of abnormal modeling of the affected femur due to alteration of the normal bio-mechanical properties of bone.

Bone scintigraphy

Exquisitely sensitive especially in polyostotic fibrous dysplasia.[1]

Complications

Malignant transformation is the most common complication.[1,3] It may be suspected due to change in the radiologic appearance of the lesion.[1] The most common malignancy is Osteosarcoma.[1,3] Other malignancies that can occur in fibrous dysplasia are fibrosarcoma and chondrosarcoma.[1] Sudden increase in alkaline phosphatase levels in one of the indicators for malignant transformation.[3] Malignant transformation is higher in males with polyostotic fibrous dysplasia, cranio-facial lesions and monostotic fibrous dysplasia.[3]

Treatment

Fibrous dysplasia is usually self-limiting except in syndromic cases which require surgery.[10] Surgical correction is an effective treatment.[2,3,8,9] Radiotherapy is avoided in fibrous dysplasia as it is radioresistant[2] and also it can induce malignant changes in fibrous dysplasia.[9]

Differential diagnosis[1,2]

- Simple bone cyst
- Giant cell tumor
- Fibroxanthoma
- Neurofibromatosis
- Hyperparathyroidism.

CONCLUSION

Fibrous dysplasia is a developmental anomaly in which normal bone marrow is replaced by fibro-osseous tissue. It is a skeletal developmental disorder of bone forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation. In our study, female predominance was noted and the disease presented in young females. All the three females presented with lesion in the cranium. The two males presented with lesions in the limb. The patients with cranial involvement did not have cutaneous lesions or peripheral bone involvement. There was no endocrinological abnormality in these patients. The study clearly stated the predominance of monostotic form over the polyostotic form.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Jagdale A, Saurav M, Patel K, Shaikh A, Prasla S. Fibrous dysplasia: a case series of five cases. Int J Adv Med 2016;3:1068-73.