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Polyosteotic Fibrous Dysplasia in a Child

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ABSTRACT:
Fibrous dysplasia is a congenital condition where normal bone is replaced with fibrous stroma and islands of immature woven bone. Polyostotic variety has early age of presentation and diagnosed usually accidentally in children below 10 years old. One such rare case was brought to us at department of radiology in which a 6 years male child was presented with complain of deformity in the left lower limb and abnormal gait which was progressive over the period of the 3 years and diagnosed as a case of the polyosteotic fibrous dysplasia on the basis of the clinical, laboratory and radio-imaging findings. The case is presented here as it is rare of its kind.

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INTRODUCTION:

Fibrous dysplasia is a congenital condition where normal bone is replaced with fibrous stroma and islands of immature woven bone. Monostotic affects single bone while polyosteotic affects multiple bones. Polyostotic has early age of presentation and usually presents at the age of 10 years old. It is painless and has no symptoms and is accidently diagnosed during investigation of another medical problem. Levels of transcription factor C-fos are raised leading to gene over expression and tumour formation. Usually bone lesions do not progress beyond puberty and so no treatment is required.

Case Presentation:

Six years male child was presented with the complains of deformity in the left lower limb and abnormal gait. This deformity was progressive over the period of the 3 years. The patient had no significant past or family history of any disease. On general examination vitals are stable. No evidence of pallor, cyanosis, or any stigmata of the neurfibromatosis. Respiratory system, cardiovascular system and neuronal examination were normal. Hepatomegaly and splenomegaly was not present.

Routine blood investigation revealed hemoglobin 12 gm/dL, total white blood cell count 6700 cells/mm^3, platelet count 2.6 lack cells/mm^3, serum creatinine 0.8 mg/dL (normal range-0.5 to 1.8 mg/dL), serum alkaline phosphate 62 IU/L (normal range-44 to 147 IU/L), serum calcium 8.8 mg/dL (normal range- 8.5 to 10.2 mg/d) and serum phosphate 2.9 mg/dL (normal range-2.4 to 4.1 mg/dL).

X-ray of the lower limb showed, ground glass appearance with the fewer cystic and sclerotic changes were evident in the left pelvis, femur, tibia, fibula and metatarsal bones. There was an over growth of the left lower limb as compare to the upper limb. No evidence of the periosteal reaction was found. Then patient was further posted to the computed tomography examination of lower limb, which showed ground glass opacities with homogeneously sclerotic area & few cystic changes in left pelvis, femur, tibia, fibula and metatarsal bones. The lesions had well-defined borders. There was expansion of the bone, with few cortical breaks seen. There was absence of the periosteal reaction.[Figure-1,2,3] In MRI of the left lower limb, lesion showed heterogeneously low signal intensity in the T1 weighted, low signal T2 weighted and

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STIR images consisting with the fibrous matrix. Few T2 and STIR high signal intensity cystic area were present. [Figure- 4, 5] Patient was diagnosed as a case of the polyosteotic fibrous dysplasia on the basis of the clinical, laboratory and radio-imaging findings. Bisphosphonate was started as supportive treatment to strengthen the bone.

DISCUSSION:

Fibrous dysplasia is a congenital condition where normal bone is replaced with fibrous stroma and islands of immature woven bone. It is a non-neoplastic tumour like process due to localised defect in osteoblastic differentiation and maturation. Fibrous dysplasia is very rare and has no cure. Fibrous dysplasia can affect any bones of body but it is commonly seen in bones of skull, ribs, shin, upper arm, pelvis and thighs. It can be divided into 4 subtypes: monostotic, polyosteotic, craniofacial fibrous dysplasia and cherubism.[1] Monostotic affects single bone, polyosteotic affects multiple bones, craniofacial fibrous dysplasia affects skull and facial bones alone and cherubism affects mandible and maxilla alone.

Fibrous dysplasia is diagnosed in childhood or early adulthood. 75% of patients is presenting before the age of 30 years. The highest incidence is

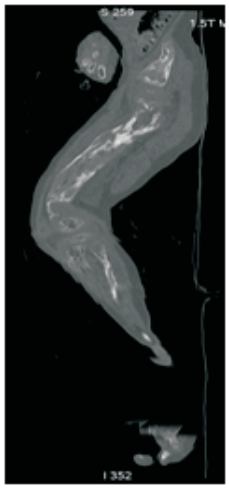

Figure 1: Sagittal CT image of left lower limb.

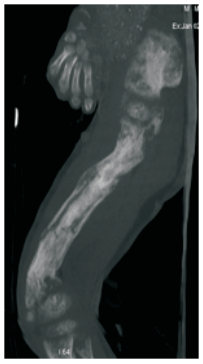

Figure 2: Sagittal MIP (Maximum Intensity Projection) CT image of left thigh in bone window shows ground glass opacities with homogeneously sclerotic area & few cystic changes in left pelvis and femur. There was expansion of the bone, with few cortical breaks seen. There was absence of the periosteal reaction seen in the age group of 3-15 years. Polyostotic has early age of presentation and usually presents at the age of 10 years old. There is no gender and race predilection.[2] As in our case of polyosteotic fibrous dysplasia was present at the age of 6 years which was consistent with these findings.

Fibrous dysplasia is a sporadic disease; still it has some associations with McCune-Albright syndrome, Mazabraud syndrome, isolated endocrinopathy without the full McCune-Albright syndrome like hyperthyroidism, hyperparathyroidism, acromegaly, diabetes mellitus, cushing syndrome and growth retardation.

In majority of the cases, it is painless and has no symptoms and is accidently diagnosed during investigation of another medical problem. In some cases it may be symptomatic and may have symptoms due to irregular bone growth and bone deformity causing compression and/or displacement of adjacent structures. This happens commonly in craniofacial fibrous dysplasia where the contents of the orbit or cranial nerves may be compressed. Bones affected due to fibrous dysplasia are weaker than normal and so susceptible to pathological fractures. Osteosarcoma, fibrosarcoma, malignant fibrous histiocytoma or rarely chondrosarcoma can be develop from fibrous dysplasia. It is more common in polyostotic form.

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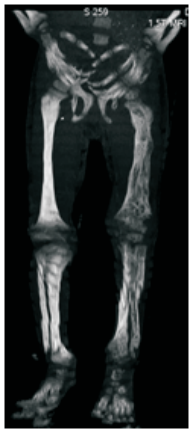

Figure 3: Coronal MIP (Maximum Intensity Projection) CT image of bilateral lower limb in bone window shows ground glass opacities with homogeneously sclerotic area & few cystic changes in left pelvis, femur, tibia, fibula and metatarsal bones. There was expansion of the bone, with few cortical breaks seen. There was absence of the periosteal reaction.

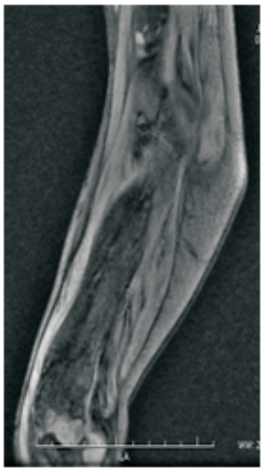

Figure 4: Sagittal T2 weighted MR image shows heterogeneously low signal intensity consisting with the fibrous matrix in femur.

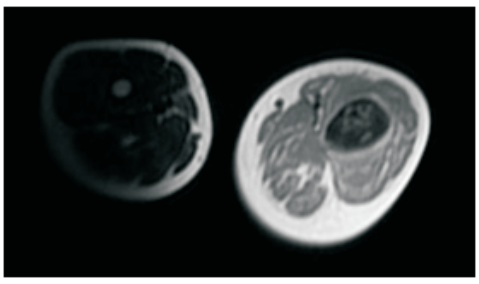

Figure 5: Axial T1 weighted MR image shows heterogeneously low signal intensity in femur.

In many cases of fibrous dysplasia levels of transcription factor is raised leading to gene over expression and tumour formation.[3] Macroscopically, medullary cavity of bones is filled with greyish-white fibrous tissue. This causes expansion of the bone involved. The bony trabeculae are thin and irregular. Many cases have Shepherd Crook's deformity which is coxa vara angulation of proximal femur. Microscopically, fibrous matrix with scattered curvilinear trabeculae of woven bone without any surrounding osteoblastic rimming is appreciated. Histologically, fibrous tissue with randomly occupied bony trabeculae are seen. The bony trabeculae are thought to be formed due to metaplasia of fibrous stroma. The fibrous stroma is of low cellularity and has variable amounts of myxoid material to dense collagenous matrix. Plump, ovoid nuceli are common, but elongated narrow nuclei can also be seen. The osseous trabeculae are made up of immature woven bone. It is not typically lined by osteoblasts. Focal osteoblastic rimming may be seen. On the concave side of the trabeculae, osteoclasts can be seen. The outline of the tropics has various shapes like solid, irregular, round islands to short, serpiginous or curvilinear shapes giving the characteristic "Chinese character" or "alphabet soup" appearance.[4]

Monostatic form mainly affects ribs, proximal femur, craniofacial bones, tibia and humerus. Polyostotic form affects femur, tibia, pelvis, foot, ribs, skull, lumbar spine, clavicle, and the cervical spine. Plain x-ray shows a ground glass matrix in the bone with completely cystic or sclerotic changes. The lesion is well circumscribed. However, periosteal reaction is absent. On CT scan, ground glass opacities, homogenously sclerotic or cystic area and expansion of bone with intact overlying cortex may be present. As the lesion is metabolically active increased uptake is seen in Tc^{99} bone scan. Lesion remains metabolically active till adulthood. MRI is not particularly useful. The lesion is heterogeneous signal; usually intermediate in T1 weighted MRI. Heterogeneous signal, usually low, in T2 weighted images. The lesion shows heterogeneous post contrast enhancement.

Differential diagnosis is Paget's disease, osteofibrous dysplasia, adamantioma and neurofibromatosis type 1. Usually bone lesions do not progress beyond puberty and so no treatment is required. Still patients can be given pain medications and medications to strengthen bones. Bisphosphonate is helpful. If there is a mass effect due to compression, surgery is considered.

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