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Monostotic Fibrous Dysplasia in Infant A Rare Presentation of Common Disease: A Case Report

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Abstract: *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. FD can be manifested as a single lesion (monostotic) or as multiple lesions (oligostotic or polyostotic). The vast majority of clinically significant bone lesions are detectable by age 10 years, with few new and almost no clinically significant bone lesions appearing after age 15 years . In this report, we describe a rare case of fibrous dysplasia in an infant. A 6 month old child presented with complains of bony swelling over right tibia. Radiological examination revealed osteolytic lesion of proximal part of right tibia. In histopathology examination, typical Chinese pattern of trabeculae along with fibrous proliferation finally diagnose case as fibrous dysplasia. Fibrous dysplasia which is a rare entity in infantile age group should be kept as possible differential diagnosis for infants presenting as painless bony swelling with no possible explanation of etiology and early operative procedure with pathological examination must be done for early diagnosis and treatment.*

Keywords: *Fibrous dysplasia, infant, monostotic, proximal tibia*

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I. Introduction

Osteofibrous dysplasia is an unusual tumour like fibro osseous lesion, originally described by Frangenheim in 1921. The other terms describing this condition are - congenital fibrous dysplasia and ossifying fibroma of bone. The term fibrous dysplasia (FD) was coined by Lichtenstein in 1938 to describe a subset of benign bone tumors manifesting in childhood or early adult life. 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. FD has a tendency of predominantly unilateral involvement and a prolonged clinical course characterized by pain, deformity and pathologic fracture of the affected bones [1]. FD can be manifested as a single lesion (monostotic) or as multiple lesions (oligostotic or polyostotic). According to Ippolito and colleagues, in monostotic FD, the most commonly appearing site is on the femur. Tibia, humerus, rib, clavicle and craniofacial skeleton are the next in order of frequency [2]. McCune-Albright syndrome is known as a combined condition consisting of polyostotic bone involvement, precocious puberty, hyperthyroidism and café au lait cutaneous macules [3,4]. Mazabraud syndrome is associated with single or multiple intramuscular myxomas in monostotic or polyostotic form [5].

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. Individual bone lesions typically manifest during the first few years of life and expand during childhood. The vast majority of clinically significant bone lesions are detectable by age 10 years, with few new and almost no clinically significant bone lesions appearing after age 15 years. In this report we describe a very rare case of 6 month old child with monostotic fibrous dysplasia involved solely in proximal tibia.

II. Case Presentation

A male child 6 month old age bought by his mother to ortho OPD with complains of swelling over right leg from 1-2 month which was progressively increasing in size and tender to touch. Swelling is not assosiated with any history of trauma or previous fracture,not assssiated with history of fever,weight loss. General physical examination revealed no pallor, icterus , cyanosis or clubbing. There is no any lymph node enlargement or peripheral edema. There is no focal or neurological deficit. Clinical examination revealed bony swelling over proximal part of right tibia with tenderness and deformity . proximal femoral and posterior tibial artery were palpable. Patient was thoroughly examined for cutaneous pigmentation or any other body patches to

rule out mc cune Albright syndrome. Radiological examination comprising x-ray of pelvis and thigh antero posterior view shows osteolytic lesion of proximal 1/3 of right tibia with well circumscribed lucent lesion with sclerotic borders in intra medullary region of proximal tibial shaft with expansion of cortex ,no periosteal reaction giving ground glass appearance, loss of normal trabecular patttern of bone was seen with surrounding soft tissue of bone appearing normal (fig 1). Chest xray was absolutely normal.

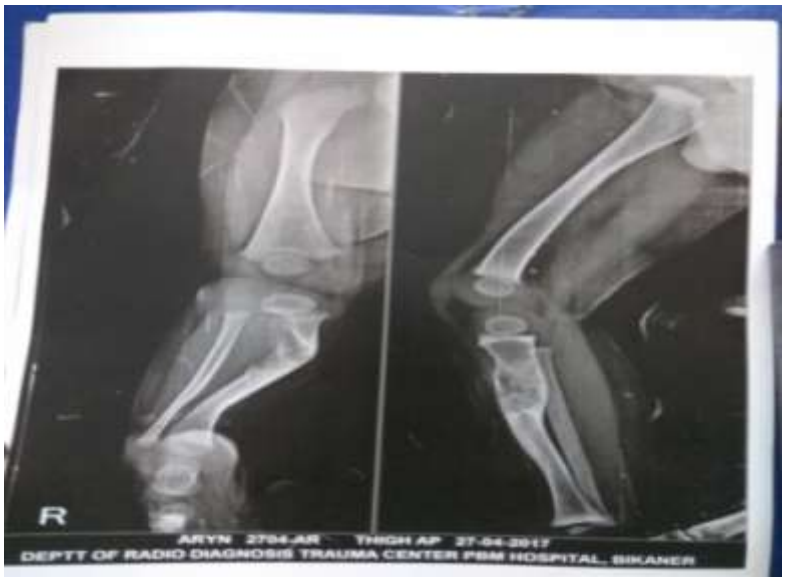

Figure 1

All laboratory test results including serum markers were with in normal range. There was no any endocrinological abnormality. On the basis of clinical and radiological finding firstly a benign bony lesion was suspected. Biopsy of osteolytic lesion of proximal 1/3 of right tibia was sent to histopathology laboratory. Macroscopically two gray white tissue piece was received which was collectively measuring 0.3x0.3cm and having smooth irregular surface. These tissue pieces were subjected to tissue processing routinely. Microscopic examination of hematoxylin and eosin stained section of osteolytic lesion of right side of tibia shows numerous delicate trabeculae arranged in different forms lacking osteoblastic rimming. Intervening spaces showed fibrous proliferation. Typical Chinese letter pattern of the trabeculae was evident. Few marrow spaces with blood vessels were present. The fibrous connective tissue was mature and well-formed bone trabeculae as shown in fig 2.1 and 2.2 below.

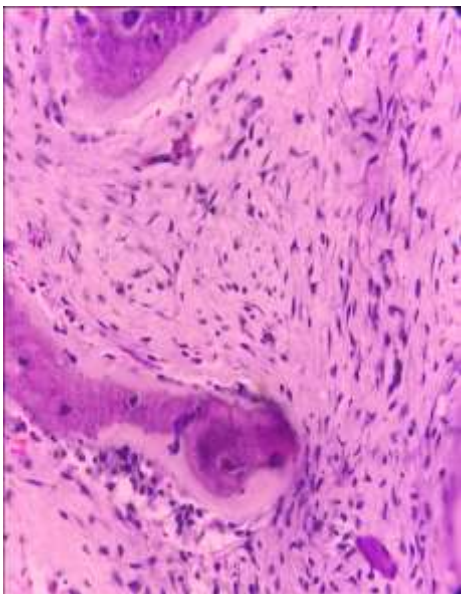

fig 2.2

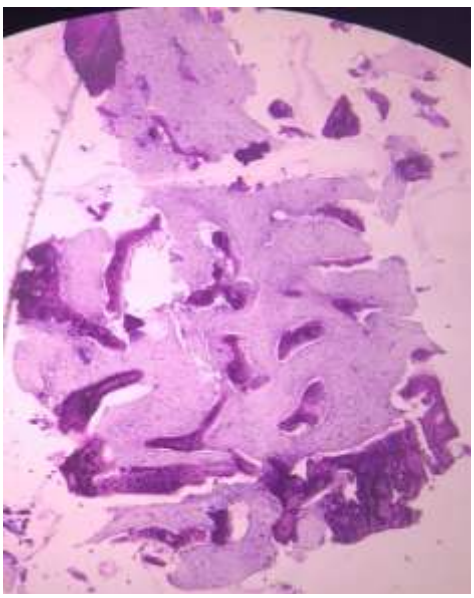

fig 2.3

III. Discussion

Fibrous dysplasia is a rare benign bone disorder characterized by the replacement of normal bone and marrow with fibrous tissue intermixed with irregular woven bone. It begins around 10 years of age and then progresses throughout adolescence. Single-bone lesions of monostotic presentation without any other disturbance are the most common forms of FD, and monostotic FDs are known to enlarge in proportion to skeletal growth[6]. Polyostotic forms are less common and often continue to enlarge after complete skeletal maturation. This feature can cause progressive deformity and increased prevalence of pathologic fractures in polyostotic FD[7]. The most common skeletal deformity in FD is a discrepancy between the lengths of bilateral limbs, referred to as shepherd's crook deformity of the proximal part of the femur.

Various theories have been proposed regarding the etiology of FD, including the trauma with a nonspecific disturbance in local bone reaction, [8] a congenital anomaly "Perverted" activity of mesenchymal bone-forming cells, [9] and a complex endocrine disturbance with local bone susceptibility. The most acceptable theory is the abnormal activity of mesenchymal cells. The pathogenesis of FD 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. When the maturation fails, it leaves behind a mass of immature isolated trabeculae enmeshed in dysplastic fibrous tissue that are turning over constantly but never completing the remodeling process. [7] The combination of insufficient mineralization and a lack of stress alignment can lead to loss of mechanical strength. This can lead to deformity, pain and pathologic fractures.

The clinical presentation varies depending on where in the cell mass the mutation is located and the size of the cell mass during embryogenesis when the mutation occurs [8].The radiographic features of FD 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. The radiographic picture is more radiolucent and well defined in the early stages and becomes mottled and more radio opaque as the disease progresses.

The histopathologic hallmark of the FD is fibrous tissue and immature, spindle-shaped, fibroblast-like cells within the bone marrow, and these fibrous tissues expand from the medullary cavity to the cortical bone. The strangely shaped trabeculae have been likened to "alphabet soup" or "Chinese characters." In our histologic results, immature bone filled with fibrous blastoma was found, and osteoblastic reaming was hardly seen. Chondroblastomas, subchondral cysts, infection-like Brodie abscesses and low-grade intramedullary central osteosarcomas can also occur in the epiphysis. Bone cysts may have more radiolucent lesions with thinner borders of lamellar bone and may show straw-colored fluid when aspirated. Chondroblastomas can be seen as subtle cartilaginous matrix on radiographs. They can present as marrow edema and joint effusion on MRI scans. Brodie abscesses can be displayed as serpiginous changes on radiographs and as the penumbra sign on MRI scans. Low-grade intramedullary central osteosarcoma is rare, but should be differentiated and may show the permeative border with a lack of a reactive shell, denser mineralization and more aggressive changes over time. Chondroblastomas, clear cell chondrosarcomas and Brodie abscesses are commonly painful[7].

IV. Conclusion

Fibrous dysplasia which is a rare entity in infantile age group should be kept as possible differential diagnosis for infants presenting as painless bony swelling with no possible explanation of etiology and early operative procedure with pathological examination must be done for early diagnosis and treatment.

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