MONOSTOTIC OSTEOFIBROUS DYSPLASIA OF THE TIBIA

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Abstract

An 11 years old boy presented with painful swelling of leg following fracture of tibia. Osteofibrous Dysplasia is an uncommon, benign, non-hereditary bone disorder in which fibrous tissue develops in place of normal bone that affects the young adults in their first and second decade of life. Tc-99m MDP whole body bone scintigraphy revealed increased tracer uptake in dense proximal tibia. On SPECT-CT radiological features of cortical based lesion superimposed on abnormal tracer uptake confirmed it to be a monostotic osteofibrous dysplasia. This case emphasizes the role of SPECT-CT and MRI in detecting osteofibrous dysplasia and differentiating it from other benign bone conditions.

Keywords: Monostotic Osteofibrous dysplasia, tibia, Bone scintigraphy, SPECT-CT, MRI

Introduction:

Osteofibrous dysplasia (OFD) is a rare, asymptomatic, nonneoplastic condition of unknown origin that affects the long bones. It has been postulated to arise from a fibrovascular defect. It has been closely associated with adamantinomas. According to one theory, OFD results from an abnormality in the haversian canals, whereas adamantinoma develops secondary to a defect of intramedullary vasculature. OFD was first described by Frangenheim in 1921 and reported it as a congenital osteitis fibrosa. Subsequently, many authors described this lesion in literature and finally, Campanacci and Laus devised the term osteofibrous dysplasia of the tibia, replacing ossifying fibroma because of its congenital origin, the histologic resemblance to fibrous dysplasia, and the dominant involvement of the tibia and fibula. Osteofibrous dysplasia is also referred to as Campanacci syndrome.

OFD is a slow growing cortical based lesion. In contrary to Fibrous dysplasia that is a medullary cavity based lesion and mainly affects the craniofacial skeleton, long bones, and ribs, OFD mainly affect the tibia or fibula and in few cases, radius or ulna. The tibial involvement was found in 35 cases reported by Campanacci and Laus; four cases had ipsilateral involvement of the fibula (11%), and 22 lesions (63%) affected the middle third of the tibial diaphysis. Ishida et al reported 11 lesions (92%) in the proximal tibial diaphysis, with one lesion in the ulna.

Osteofibrous dysplasia is usually diagnosed in children younger than 10 years, with a peak incidence in children aged 1-5 years. Several newborn cases have also been reported. Since OFD is a rare disease, there is limited data in the literature about characteristics of disease and response to medical treatment. There is no spontaneous resolution of OFD. Non-surgical treatment is recommended until skeletal maturity is reached. Recurrent pathologic fractures may be an ongoing problem in some active children. Using a tibial brace similar to those used for congenital pseudarthrosis of the tibia may minimize recurrent pathologic fractures. Surgery should be performed if complications are imminent, such as marked bone deformation or pseudoarthrosis.

Case Report:

An 11 years old male boy presented with a painful, progressively increasing swelling of left tibia for a
few months. Preceding this complaint, there was a history of fracture at the same site 4 years back while playing and was treated with cast. No associated history of weight loss, fever or night sweats. On examination, there was a tender swelling involving the extensor surface of left proximal leg with no signs of inflammation. Distal neurovascular examination was unremarkable. Three Phase Tc-99m Whole Body Bone scintigraphy (Figure 1) revealed hyperemia, increased blood pool activity and abnormal intense tracer uptake involving proximal 1/3 of diaphysis of left tibia. Additionally, subtle focal tracer uptake was seen in the mid diaphysis of left tibia. On SPECT-CT images (Figure 2), there was a corresponding expansile, predominantly lytic, cortical based, “ground glass” appearance involving proximal left tibial diaphysis with peripheral zone of sclerosis and cortical thickening. There was no associated soft tissue component. A green stick pathological fracture line was also seen adjacent to this lesion. Faint tracer uptake in mid tibia showed underlying lucenty with peripheral sclerosis. Further correlative radiological imaging [X-ray (Figure 3) and MRI (Figure 4)] supported the diagnosis of fibrous dysplasia.

Discussion:

Osteofibrous dysplasia is an unusual developmental condition of childhood, which mainly affects the tibial diaphysis. It follows a slowly progressive course and tends to stabilize after skeletal maturity. The possible link with adamantinoma still remains controversial and some authors believe that they are a spectrum of same histological process.\cite{12} Histologically, OFD is characterized by an osteofibrous stroma replacing the normal bone, showing trabecular pattern which, unlike in FD, are surrounded by a regular rim of prominent cubic osteoblasts, mature lamellar bone and zonal segmentation. Radiologic features are eccentric, lytic lesions, usually involving tibial diaphyses with “ground glass” appearance. Differential diagnosis includes FD and adamantinoma. Just like OFD, the latter occurs predominantly in the tibia, its malignant potential remains high, calling for radical surgical treatment. According to the literature review, the mean age for OFD was nine years, which is considerably lower than that of patients with FD (30 years). The lesions were found exclusively in the tibia. Unlike in FD, once bone maturation is completed, surgical treatment of OFD is not normally followed by recurrent disease. Thus, the prognosis of OFD with restricted surgical therapy is more favourable than that of FD.\cite{11,12}

Scintigraphically OFD lesions show increased uptake due to high bone turnover. Although BS is sensitive for OFD, it lacks specificity. The poor specificity is improved with the use of SPECT-CT which concomitantly characterizes the metabolic and anatomic features of the disease.\cite{13} The differentiating feature from malignant conditions, like adamantinomas, is the absence of soft tissue component with a well preserved cortical margin.\cite{14} Very few case reports are published regarding use of Bone SPECT-CT in detection of Osteofibrous dysplasia.\cite{15} Radiologic correlative imaging like MRI and X-rays are further helpful in characterization and diagnosis.

**Fig. 1:** Three Phase Bone scintigraphy (A) showed hyperemia and increased blood pool activity in the proximal left leg (black arrows). Delayed static images (B) show hot spots in the left tibial diaphysis caused by abnormal increased uptake of the radiisotope technetium-99m methylene diphosphonate (99m Tc MDP).
Figure 2: SPECT/CT images showed MDP avid expansile cortical lesions involving left proximal tibial diaphysis with central ground glass appearance and peripheral intact zone of cortical thickening.

Figure 3: Cranio-caudal (CC) view; ill-defined radio opaque mass in left retroaerolar region, protruding on skin.

Figure 4: Magnetic resonance imaging (MRI) shows typical features with low to intermediate signal intensity equal to that of muscle on T1-weighted image (A). Nodular enhancement pattern is seen on post Gadolinium contrast enhancement sequence (B). No surrounding medullary edema is seen on STIR contrast image (C).

References: