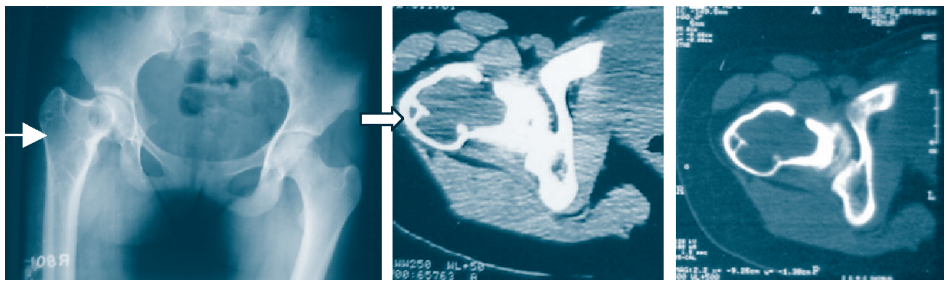
Normal technetium-99m-MDP uptake in fibrous dysplasia of the hip

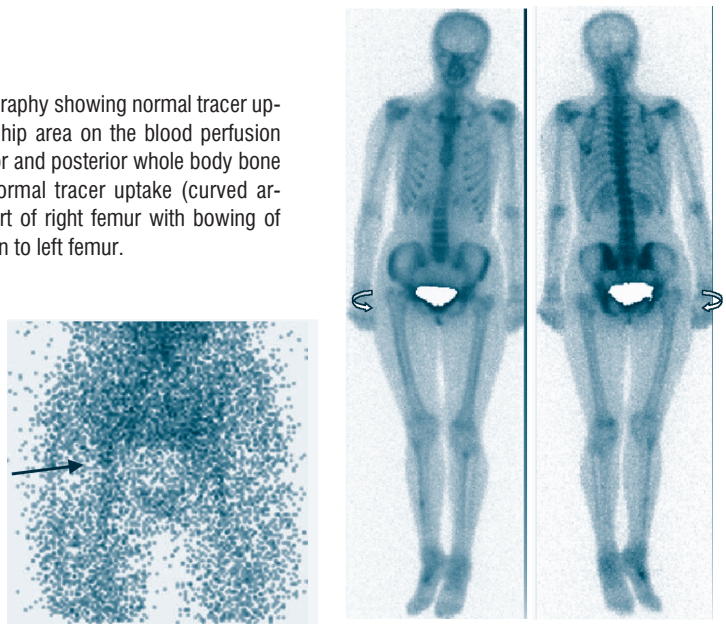
To the Editor: We would like to describe an entity not published in HJNM so far: Fibrous dysplasia is a congenital, sporadic developmental disorder of the skeletal system. It is idiopathic in nature and occurs in 10.4%-46.5% according to different studies [1]. Osteoblasts fail to undergo normal morphological differentiation and maturation. The medullary space is replaced by fibro-osseous tissue resulting in an expansile remodeling of the affected bone. Lichtenstein demonstrated fibrous dysplasia as a distinct entity in 1938 and it is also known as Lichtenstein-Jaffe disease [1]. The lesion may be surrounded by a layer of thick sclerotic reactive bone termed as rind [1, 2]. Most of the patients are diagnosed in the 1st or the 2nd decade of life though it has also been diagnosed in later life, with slight female predilection [1]. Most of the patients are asymptomatic and the lesions are found incidentally on unrelated diagnostic workup. Some may present with pain or a mass [1]. Femur, ribs and facial bones are frequently involved [3]. Fibrous dysplasia results in deformities like leg length discrepancy (70%), shepherd's crook deformity of the proximal femur (35%), facial asymmetry, tibial bowing and rib deformity [4]. Most of the fibrous dysplasias are monostotic in nature (70%-80%) while the rest are polyostotic [5]. Some of the patients (36%) may show increased levels of serum alkaline phosphatase [6].

We have studied a 17 years old female with a few non-progressive symptoms in the right thigh. There was no history of trauma or fever. The physical examination and laboratory test were within normal limits. There was no periosteal reaction in computerized tomography (CT) images and in the X-ray films. The right proximal femur was deformed with reduction of the angle between the neck and shaft leading to coxa vara and shepherd's crook deformity (the bowed appearance). CT scan showed soft tissue attenuation (40-55 Hounsfield units) in the soft tissue window settings (Fig. 1A, B and C). There was a break in the anterior cortex of the neck of the right femur with smooth margins. No significant soft tissue swelling or periosteal reaction was noted. There was no ground glass haze. The 3 phase bone scintigraphy was performed with technetium-99m-methylene diphosphonate (^{99m}Tc-MDP) (Fig. 2A and B). The blood perfusion phase, acquired soon after injection of bone scintigraphy of the hips revealed normal tracer concentration on the affected right side. The blood pool phase acquired after 5min of injection was also normal on both sides of hip. The delayed bone scan, performed after 3 hours of injection, revealed normal tracer uptake in the proximal part of the right femur. Usually this uptake is markedly increased due to increased vascularity. Histopathology revealed fibrous dysplasia (Fig.3). We decided to keep the patient under observation. Characteristic radiological findings that support the diagnosis of fibrous dysplasia are: well defined osteolytic lesions surrounded by zone of sclerosis, deformation, expansion with cortical thinning and over all ground glass appearance [7].



A B C

Figure 1. A: AP radiograph of hip joints shows a multiseptal geographic lytic lesion (arrow) in proximal metadiaphysis, neck and greater trochanter of the right femur. Coxa vara and shepherd's crook deformity are also noted. B: CT section in bone window settings shows trabeculated lytic lesion with rind of sclerosis. (arrow) C: CT section in soft tissue window settings shows replacement of bone marrow fat by soft tissue (40-55HU) (thick arrow).



A B

Figure 2. A: Bone scinitigraphy showing normal tracer uptake (arrow) in the right hip area on the blood perfusion phase. B: Delayed anterior and posterior whole body bone scinitigraphy showing normal tracer uptake (curved arrows) in the proximal part of right femur with bowing of femur shaft in comparison to left femur.

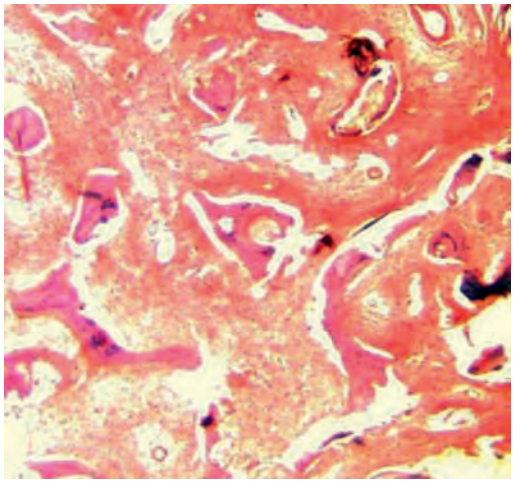


Figure 3. Histological slide of fibrous dysplasia, 400x, (H-E) stain. M/E shows fibrous tissue of variable cellularity interspersed with bony trabeculae in a Chinese letter pattern.

However conditions like chondroblastoma, enchondroma, solitary bone cyst, giant cell tumour, nonossifying fibroma, aneurysmal bone cyst, bone infarction and chronic bone abscess need to be differentiated [2, 5, 7-10]. Fibrous dysplasia rarely undergoes malignant transformation [8].

The absence of increased tracer uptake does not mean that the diagnosis should be excluded [10-16]. The bony conditions like osteoid osteoma, aneurysmal bone cyst, chondroblastoma, osteoblastoma and giant cell tumour show increased tracer uptake while bone cyst, bone island, cortical desmoids, enchondroma, fibrous cortical defect and intraosseous ganglion show normal to mildly increased tracer uptake on bone scintigraphy [11]. The interest of our case report lies on the following: a) Normal uptake in fibrous dysplasia is seldom seen, b) Bone scintigraphy alone should not be taken as an exclusion criterion for fibrous dysplasia.

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