

Late diagnosed polyostotic fibrous dysplasia. Bone scan, radiography and magnetic resonance imaging findings

To the Editor: We read with interest a case of fibrous dysplasia (FD) published in Hell J Nucl Med. 2009; 12(1):72-73 [1] and we present another case which differs to the above as having multiple bone involvement on bone scintigraphy. The patient was a 12 years old girl with right hip joint pain. When she was six years old following a fall she had a right femoral neck fracture and underwent reparative surgery. No imaging study other than plain radiography was done at that time and no definite diagnosis was made. Since then she has been suffering a progressive right hip pain and limping on the same side. There was a significant discrepancy in the length of the lower extremities and a facial deformity as right mandible protrusion and hypertelorism. No skin lesions or pubertal disturbances were noted. Serum alkaline phosphatase was elevated, but serum calcium and phosphorus were within normal limits. Other related laboratory tests were normal.

Whole body bone scintigraphy was performed after the intravenous injection of 740MBq ^{99m}Tc -methylene diphosphonate (^{99m}Tc -MDP). The planar whole body images were obtained three hours after the injection and showed multiple regions of intense activity in the right side of the skull, right side of the mandible, right humerus, right scapula, right hemi pelvis, right femur and right tibia. Foci of increased uptake were also noted in the lumbar spine and the left humerus (Fig. 1).

Plain radiography of pelvic bones showed radiolucent areas of expanded bone with a ground-glass pattern in the proximal right femur. The typical "shepherd's crook" deformity was visible on this side. The radiolucent regions were also present at the right pelvis (Fig. 2).

The corresponding magnetic resonance imaging (MRI) of the lower extremities demonstrated in the T1-weighted coronal images, hypo-intense lesions in the proximal right femur. Bowing deformity of the right femur was also noticed (Fig. 3).

Fibrous dysplasia is a developmental bone disorder in which normal bone is replaced by fibroblasts and immature woven bone [2, 3] occurs equally in men and women and is most common in the first two decades of life [3-5]. Endocrinopathies occur in 2%-3% and include hyperthyroidism, hyperparathyroidism, acromegaly, diabetes mellitus, and Cushing's syndrome [6].

Monostotic type of FD which involves one bone accounts for the majority of cases (80%). This type



Figure 1. The whole body images show multiple regions of intense activity on the right side of the skull, mandible, right humerus, scapula, hemi pelvis, femur and tibia. Foci of increased uptake were also noted in the lumbar spine and the left humerus.



Figure 2. Plain radiography of the pelvic bones shows radiolucent areas of expanded bone with a ground-glass pattern in the proximal right femur. The typical "shepherd's crook" deformity is visible on this side. The radiolucent regions are also present in the right pelvis.

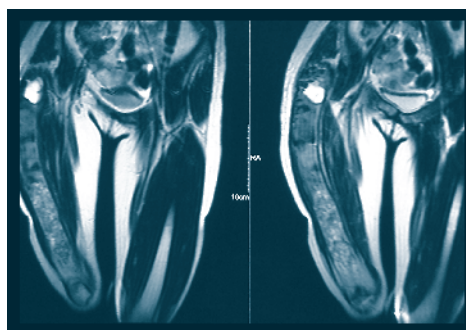


Figure 3. The corresponding MRI of the lower extremities demonstrates hypo-intense lesions on the proximal right femur in T1-weighted coronal images. Note also the bowing deformity of the right femur.

may be asymptomatic and diagnosed incidentally while the polyostotic fibrous dysplasia (PFD) is often more severe [2, 7]. Approximately 2%-9% of the general population [1] and 15% of all patients with FD [8] have PFD. Nearly two thirds of PFD patients will be symptomatic before the age of 10 [9].

The association of PFD, precocious puberty and café-au-lait spots, "McCune-Albright syndrome", is the most severe form of the

disease [10]. Most common symptoms are bone pain, fractures and bone deformities. Neurological deficits depending on the region of involving skull may occur in these cases [10, 11]. Subjects with involvement of orbital bones, may suffer from hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos, and blindness. Subjects with temporal bone involvement may suffer from vestibular dysfunction, tinnitus, and hearing loss. Olfactory disorders may result from cribriform plate involvement [10, 11]. In PFD, multiple bones unilaterally or bilaterally are affected [4, 5, 11]. They frequently involve the femur, tibia, skull, ribs, pelvis, facial bones, and less often the upper extremities, clavicle, lumbar and cervical spine. Spine involvement is rare and almost always occurs only in PFD. Limping with a low grade pain or pathologic fractures are common in PFD [7]. Pathologic fractures are due to the osteolytic lesions weakening the long bones or to deformities which impair the mechanical tolerance of the bones [10]. There is also a risk of malignant transformation in the PFD type of the disease [7]. In craniofacial bone involvement, face asymmetry and swelling are usually seen [10].

Bone scintigraphy reveals intensely increased radiotracer uptake in the affected bones [11, 12] but is not routinely used as a diagnostic tool [7]; however it is useful to diagnose the extent of the disease [12]. Characteristic lesions of FD on bone scanning such as bar-shaped pattern, whole bone involvement, and a close match between the size of the lesion on radiographs and bone scanning are helpful to differentiate FD from other diseases [12]. The differential diagnoses of PFD are multiple enchondromatosis, primary hyperparathyroidism, neurofibromatosis, multiple bone hemangiomas, eosinophilic granuloma, fractures, Paget's disease and osteoblastic metastases [1, 6, 13]. Diagnosis may be facilitated by pathological fractures in 85%, leg-length discrepancy in 70% and skin pigmentation in 50% of PFD cases [6]. The only noteworthy laboratory finding is an elevated alkaline phosphatase level [6].

In plain radiography in PFD, normal bone is replaced by a radiolucent tissue with a "ground-glass" pattern. The lesion was surrounded by a distinct rim. The diameter of the bone is increased and no periosteal reaction occurs [7], [12]. The location of lesions in the long bones are usually diaphyseal or diamephyseal. The typical "shepherd's crook" is a bowing deformity of proximal femur with a varus angulation [7].

In MRI FD has highly variable presentations. In T1-weighted images the lesions are often isointense to muscular tissues and in T2-weighted, they are mostly hyper-intense [7].

In conclusion, although bone scintigraphy does not play an initial role in the diagnosis of PFD, it is useful to show the

extent of the disease as in this case, and to early start treatment, that unfortunately did not happen in our case.

Bibliography

1. Sood A, Raman R, Jhobta A et al. Normal technetium-99m-MDP uptake in fibrous dysplasia of the hip. *Hell J Nucl Med* 2009; 12: 72-3.
2. Song JJ, Jung HH, Lee HM, Hwang SJ. Monostotic fibrous dysplasia of temporal bone: report of two cases and review of its characteristics. *Acta Otolaryngol* 2005; 125: 1126-9.
3. Al-Ismail K, Torreggiani WC, Munk PL, Lee MJ. Musculoskeletal case 16. Fibrous dysplasia. *Can J Surg* 2001; 44: 170 & 227.
4. Hempel JM, Karkos PD, Issing WJ. Fibrous dysplasia of the frontal bone. *Ear Nose Throat J* 2006; 85: 654, 656-7.
5. Nelson BL, Thompson LD. Fibrous dysplasia of bone. *Ear Nose Throat J* 2003; 82: 259.
6. Lichenstein L, Jaffe HL. Fibrous dysplasia of bone: a condition affecting one, several or many bones, the graver cases of which may present abnormal pigmentation of skin, premature sexual development, hyperthyroidism or still other extraskelatal abnormalities. *Arch Pathol* 1942; 33: 777.
7. Cento EA, Lomasney LM, Demos TC et al. Radiologic case study. Monostotic fibrous dysplasia. *Orthopedics* 2007; 30: 82, 166-70.
8. Rudenko B, Klingelschmitt S, Aubry S. Hemibody uptake on bone scintigraphy in polyostotic fibrous dysplasia. *Clin Nucl Med* 2003; 28: 992-3.
9. Harris WH, Dudley HR, Jr, Barry RJ. The natural history of fibrous dysplasia. An orthopaedic, pathological, and roentgenographic study. *J Bone Joint Surg Am* 1962; 44-A: 207-33.
10. Chapurlat RD, Orcel P. Fibrous dysplasia of bone and McCune-Albright syndrome. *Best Pract Res Clin Rheumatol* 2008; 22: 55-69.
11. Schoenau E, Rauch F. Fibrous dysplasia. *Horm Res.* 2002; 57 Suppl 2: 79-82.
12. Di Caprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. *J Bone Joint Surg Am* 2005; 87: 1848-64.
13. Johns WD, Gupta SM, Kayani N. Scintigraphic evaluation of polyostotic fibrous dysplasia. *Clin Nucl Med* 1987; 12: 627-31.

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