Case Report

An interesting case of polyostotic fibrous dysplasia: The "pirate sign" evaluated with Tc-99m methylene diphosphonate single-photon emission computed tomography/computerized tomography

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ABSTRACT

Polyostotic fibrous dysplasia is a rare progressive benign disorder of the bone. Bone scintigraphy is extremely useful in the initial evaluation for identifying the extent of disease. We report a case presenting with pathological fracture of the shaft of the right femur. After treatment of the fracture, bone scintigraphy revealed involvement of multiple bones including the skull and facial bones. The utility of single-photon emission computed tomography/computerized tomography in the evaluation of the extent of skull base involvement is highlighted.

Keywords: Polyostotic fibrous dysplasia, scintigraphy, single-photon emission computed tomography/computerized tomography, Tc-99m methylene diphosphonate

INTRODUCTION

Fibrous dysplasia (FD) is a benign intramedullary fibro-osseous lesion. FD may occur in one bone (monostotic) or multiple bones (polyostotic) and may be associated with other conditions like McCune Albright syndrome. Clinical presentation may occur at any age, with the majority of lesions being detected by the age of 30 years. The disease has no gender predilection. Common sites of skeletal involvement are the long bones, ribs, craniofacial bones, and the pelvis. Involvement of the sphenoid wing is commonly described as the "pirate sign".

CASE REPORT

A 32-year-old female presented with pathological fracture of the shaft of the right femur. Radiological evaluation revealed features of FD (with fracture of the right femoral shaft), with similar lesions in the opposite femur. The right femoral fracture was treated with intramedullary nailing, and intraoperative biopsy confirmed the diagnosis of FD. In order to document the extent of bony involvement in the rest of the skeleton, whole body bone scintigraphy was performed 3 hours after intravenous injection of 99mTc-Methylene diphosphonate (MDP). Multiple sites of increased skeletal tracer uptake were detected in the skull, left mandible, multiple ribs, pelvis and both lower limbs [Figure 1]. Single-photon emission computed tomography/computerized tomography (SPECT/CT) of the skull was performed for evaluation of the extent of skull bone involvement and showed involvement of the right sphenoid, ethmoid and frontal bones and left mandible, with evidence of bone expansion [Figure 2]. CT component of SPECT-CT [Figure 3] shows bony expansion with ground glass appearance involving right sided ribs. Involvement of the sphenoid wing (commonly described as the "pirate sign") was also better depicted with SPECT/CT.

DISCUSSION

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.[1] Failure of maturation leaves a mass of immature isolated trabeculae enmeshed in dysplastic fibrous tissue that constantly turns over but never (or very, very slowly) completes

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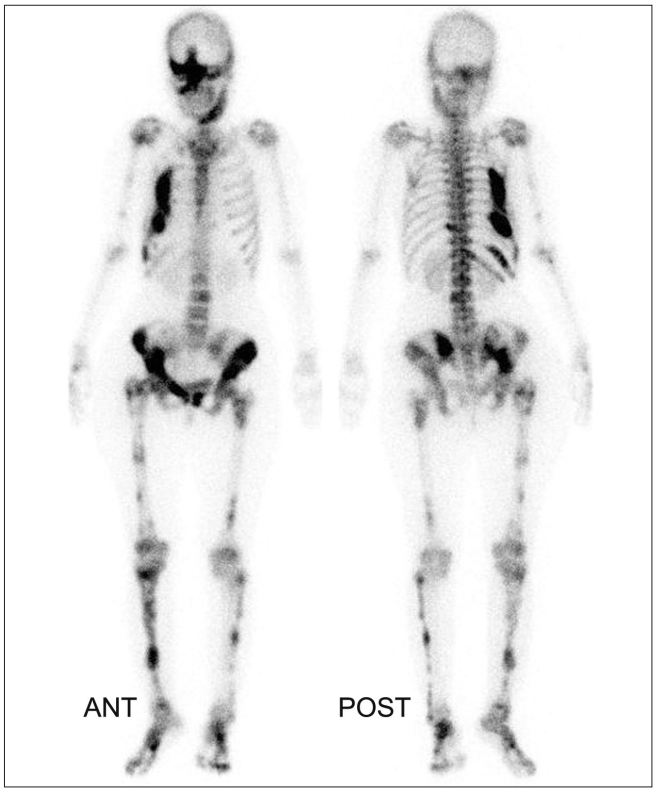

Figure 1: Tc-99m MDP whole body images show multiple sites of increased tracer uptake, including the skull and facial bones, multiple ribs, pelvis and bones of both lower limbs. The "pirate sign" indicates involvement of the right sphenoid wing

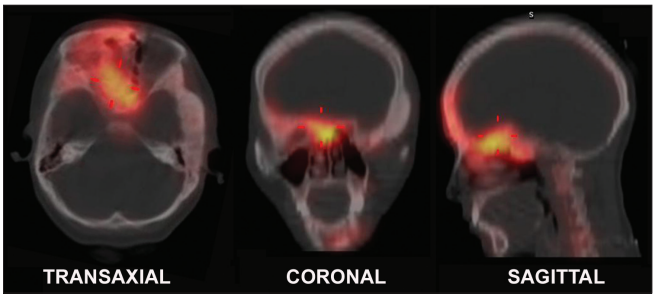

Figure 2: Hybrid SPECT/CT fusion to determine the extent of skull and facial bone involvement shows intense tracer uptake in the sphenoid, ethmoid and frontal bones

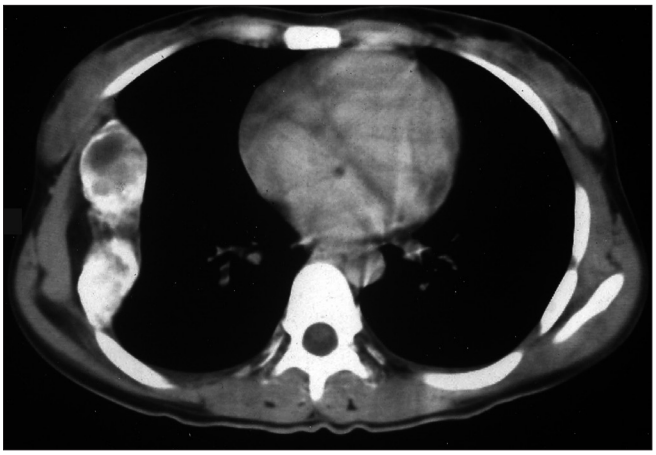

Figure 3: Non-contrast CT scan of the chest shows bony expansion and deformity of the right ribs, where intense osteoblastic activity was detected

the remodeling process. The combination of a lack of stress alignment and insufficient mineralization results in substantial loss of mechanical strength, leading to the development of pain, deformity, and pathologic fractures. The etiology has been linked with a mutation in the Gs alpha gene that occurs after fertilization in somatic cells[2] and is located at chromosome 20q13.2-13.3.

The monostotic presentation of FD is more frequent than polyostotic presentation, and lesions enlarge in proportion to skeletal growth. Polyostotic lesions often continue to enlarge after skeletal maturity, with progressive deformity and an increase in pathologic fractures. The prevalence of polyostotic FD is difficult to characterize.[3] Patients with polyostotic disease and large, painful lesions in weight-bearing long bones are at the greatest risk for pathologic fracture. Local expansion of FD in the maxilla, zygomatic, or ethmoid bones of the face can produce substantial functional and cosmetic deformity.[4] At the initial presentation, radionuclide bone scintigraphy is useful to demonstrate the extent of the disease.[5] Actively forming lesions in adolescents have greatly increased isotope uptake that corresponds closely to the radiographic extent of the lesion. Polyostotic FD has been demonstrated using F18-Fluoride positron emission tomography/computed tomography (PET/CT). The utility of volume rendering has also been demonstrated in evaluation of the bony deformities.[6] The "pirate sign" has been previously described on planar bone scintigraphy in FD.[7] Recent studies have described the advantage of performing bone SPECT in addition to CT to determine the extent of FD in the skull base and of hybrid SPECT/CT imaging in FD of the lumbar vertebra.[8,9] The present case illustrates the utility of hybrid SPECT/CT in documenting the complete extent of bony involvement of the skull base.

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