

A Case of Polyostotic Fibrous Dysplasia Masquerading as a Multiple Bone Metastases

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Abstract

Fibrous dysplasia is a benign, bony abnormality that is usually asymptomatic. A 41-year-old male with minimal symptoms presented at this hospital with abnormal findings incidentally seen in his ribs on the chest radiograph. A skeletal survey showed numerous, osteolytic lesions throughout multiple bones. Diagnostic processes for malignancy of undefined primary origin (MUO) were performed in order to identify the underlying primary neoplasm, although abnormal findings were not seen except for multiple bone lesions. A computed tomography guided bone biopsy was performed on his left rib. The final diagnosis was fibrous dysplasia. This case demonstrates that fibrous dysplasia should be considered in the differential diagnosis in young patients with multiple, osteolytic lesions and without a prior history suggesting malignancy.

Key Words : Bone neoplasm, Fibrous dysplasia, Metastasis

Introduction

Fibrous dysplasia (FD) is a rare, sporadically occurring bone disease, characterized by abnormal bone growth where normal bone is replaced by fibrous skeletal tissue composed of bone-forming mesenchyme [1]. FD represents abnormal growth and swelling of bone results poor mechanical

strength which can lead to pathologic fractures.

Most patients have lesions involved in one bone(monostotic) and others have them in multiple bone(polyostotic) [2].

The majority of monostotic lesions are asymptomatic and are discovered when radiographs of the involved region are obtained incidentally or during other intentions [3]. On the

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other hand, polyostotic forms may sometimes be accompanied by bone pain and deformities. The typical radiologic description is the “ground-glass” pattern of the bone, and which is helpful in a correct diagnosis. However, in some patients, it is difficult to differentiate the radiologic diagnosis between FD and malignant bone diseases.

We report a case of polyostotic FD with minimal symptoms masquerading as multiple bone metastases.

Case Report

In February 2013, a 41-year-old male visited to this hospital with abnormal findings on his chest radiograph taken during his annual health screening. He had suffered from fatigue and mild low back pain for one year. He was otherwise in good health and no specific past medical or family history. His physical examination results were unremarkable. His chest radiograph revealed several, osteolytic bulging masses in his left ribs (Fig. 1). His full blood count and biochemistry showed almost normal except for a mild elevation in alkaline phosphatase (228 IU/L). He was admitted for further evaluation and got an examination processes which usually used for diagnosing malignancies of unknown origin (MUO). The level of serum alpha-fetoprotein (AFP) and β -subunit of human chorionic gonadotropin (β -hCG) were normal. A skeletal survey showed multiple, osteosclerotic, bulging lesions in his skull, both ribs, both femurs, the right tibia, both humeri, and the right radius. Serum and urine electrophoresis were done and there were no abnormal peaks. The esophagogastroduodenoscopy showed short segment of Barrett’s esophagus without any other abnormalities. Computed tomography (CT) of his chest, abdomen, and pelvis

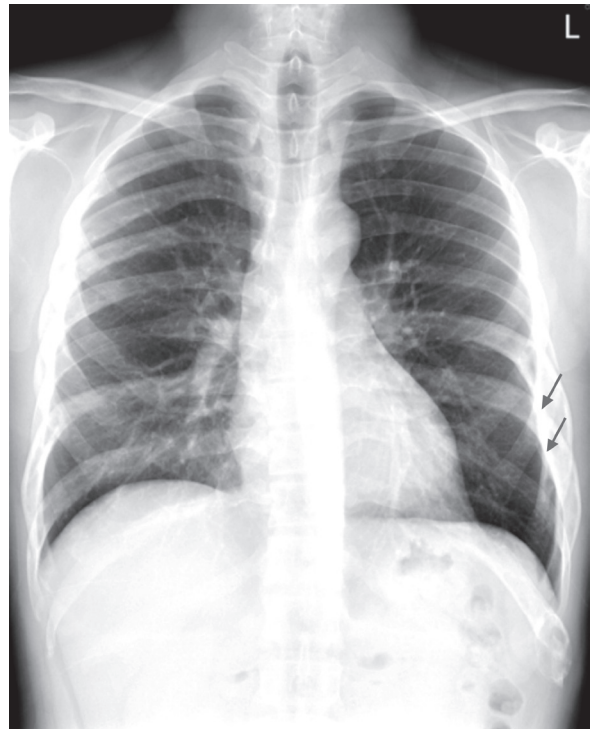


Fig. 1. Initial chest radiograph.

revealed multiple skeletal lesions in the right ischium, both femurs, cervical spines, and both side of ribs. In his abdominopelvic CT showed a 2,7-cm nodule in the right adrenal gland, but there were no other abnormal findings. On the delayed phase of enhanced CT, the washout ratio was 71%, and which met the radiologic criteria for the diagnosis of a lipid-poor, adrenal adenoma [4]. The bone scan showed multiple abnormal areas of radio-uptakes, which suggested the lesion might be metastasis. But, the scan showed diffusely increased uptakes in his right tibia, including the metaphysis and diaphysis, these findings were somewhat different from that of metastasis. The skull base was also involved, but the spine was spared in the scan, which is also unusual in malignant bone metastasis (Fig. 2). We could not find any evidence of malignancy. All results



Fig. 2. Initial bone scan.

suggested a benign bone disease, such as fibrous dysplasia or Paget's disease, rather than metastatic bone tumor. The fluorodeoxyglucose-positron emission (FDG-PET)/CT scan showed increased FDG uptake in multiple, skeletal lesions [maximum standardized uptake value (SUVmax)=12.2 g/mL in the left, 11th rib], thus favoring metastatic lesions rather than benign bone tumors (Fig. 3). However, patient's FDG-PET/CT showed only bony lesion and no other evidence of the primary site of solid malignancies. The patient underwent percutaneous, CT-guided, needle biopsy of the left, 11th rib, and three cores were obtained. Microscopically, the pathology specimens revealed a benign, fibro-osseous lesion, thus confirming the diagnosis of fibrous dysplasia. Cellular atypia, mitotic figures, or necrosis were not observed (Fig. 4).

The patient then received 60 mg of intravenous pamidronate every three months and regularly followed in the outpatient clinic.

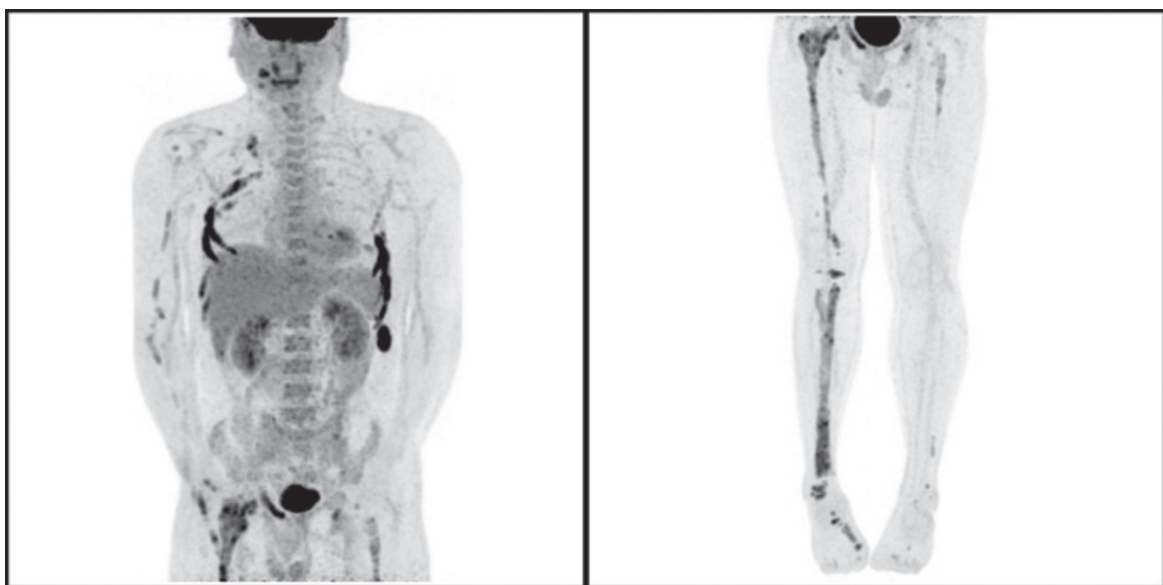


Fig. 3. FDG-PET/CT scan.

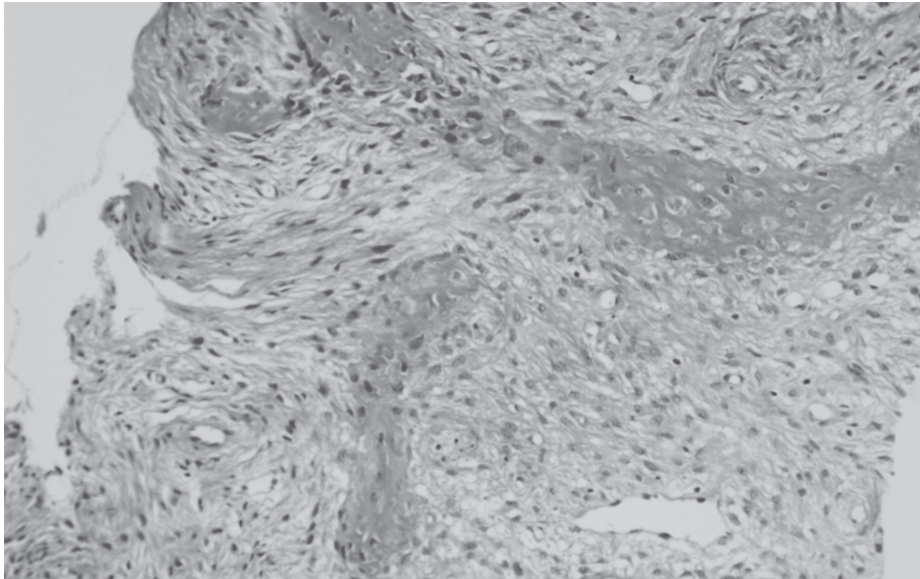


Fig. 4. Microscopic pathology.

Discussion

FD was described by Lichtenstein [5] in 1938, and the incidence and prevalence of FD are difficult to estimate, these lesions are not so rare. FD accounts for about 5% to 7% of benign bone tumors and consists nearly 1% of primary bone tumors [6].

Most lesions are monostotic, and approximately 20% to 30% of FD cases are polyostotic. The majority of monostotic lesions are asymptomatic, although polyostotic cases may be accompanied by bone pain or deformities, and two thirds of these patients are symptomatic before the 10 years of age [2].

Sometimes it is difficult to differentiate polyostotic FD from disseminated bone metastasis. The clinical and radiologic characteristics of polyostotic FD and bone metastasis are very similar in the occurrence of multiple lesions and location of, anatomic sites, but the prognosis and methods of treatment are totally different.

CT is the most successful technique for demonstrating the typical radiographic findings of FD. The CT findings are intramedullary, expansile, well-defined lesions with intact cortex and a characteristic ground-glass appearance, although some cases may appear almost radiolucent or sclerotic [7]. In this patient, the radiologic findings showed multiple, bulging lesions, and which suggested bone metastasis or multiple myeloma. However, his CT images did not show a periosteal reaction, cortical destruction or a soft-tissue mass lesions. Therefore, the CT images suggested a benign condition rather than malignancy.

Although radionuclide bone scintigraphy is non-specific, these scans are helpful in making a more reliable diagnosis. Many benign bone lesions, such as eosinophilic granulomas, disseminated tuberculosis, fibrous dysplasia, and enchondroma, appear similar to bone metastasis on bone scans [8,9]. However, there are some useful factors which differentiate FD from metastasis. First, the involved FD sites are usually extremities and ribs,

unlike metastatic bone tumors which favor the axial skeleton, especially the spine and pelvis. Second, the hot uptakes usually appear in the entire parts of the involved bone in the FD [10].

With regard to FDG-PET/CT, it is known that there is a large variability in the FDG avidity of FD. The SUVmax values of the FD have ranged from 3.73 to 9.64 in previous reports [11]. But in this case, the FDG-PET/CT scan showed higher FDG uptake in the left 11th rib. (SUVmax = 12.2 g/mL), therefore the possibility of FD should not be excluded on the basis of FDG avidity.

Radiologic findings are usually sufficient for making a diagnosis of FD [10], and a bone biopsy is not necessary. The bone biopsy leads likely to result in a fracture which can be difficult to heal [12]. Therefore, bone biopsy should be avoided whenever possible, and the diagnosis of FD should be based on the clinical and radiological findings. In this patient, objective evidence of malignancy based on the clinical, laboratory, and radiologic findings were insufficient. A potentially harmful diagnostic approach, such as bone biopsy, had been chosen due to the patient or clinician's over concern, was inappropriate.

Conclusion

Polyostotic FD may be misdiagnosed as metastatic bone disease, and FD should be included in the differential diagnosis in young patients with multiple, osteolytic lesions but who do not have a prior history of malignancy.

Conflict of Interest

The authors report no conflict of interest in this work.

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