Case Report

Paget's disease diagnosed on bone scintigraphy: Case report and literature review

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ABSTRACT

Paget's disease of bone is a chronic bone remodeling disorder. Although most patients are asymptomatic, a variety of symptoms and complications may develop directly from bone involvement or secondarily due to compression by the expanded bone. It is usually diagnosed from radiological and biochemical abnormalities or in advanced cases it becomes clinically evident due to the expanded bone. We report a case of Paget's disease which was detected incidentally during evaluation of nephrolithiasis and polyarthritis but had normal radiographs and normal biochemical markers.

Keywords: Alkaline phosphatase, bone scintigraphy, nephrolithiasis, Paget's disease

INTRODUCTION

Paget's disease of bone is a chronic bone remodeling disorder featuring one or more areas of aggressive osteoclast-mediated bone resorption, followed by imperfect osteoblast-mediated bone repair.[1] Although it is usually asymptomatic and diagnosed incidentally, most clinical manifestations are skeletal.[2] The disease is frequently recognized because of elevation in serum alkaline phosphatase levels, which reflects both the extent and the activity of the disease.[3] However, alkaline phosphatase levels can be normal when there is only a small focus of Paget's disease or after successful medical treatment.[4] We report one such case that was incidentally detected during evaluation of nephrolithiasis and polyarthralgia and had normal radiographs and normal serum alkaline phosphatase levels.

CASE REPORT

A 46-year-old man, driver by occupation, presented with a history of sudden onset pain abdomen, which was colicky in nature and localized to the right lumbar region. There was no history of dysurea, hematuria, fever, loose stools, or vomiting. He also had a history of multiple joint pains for the last 4 months, aggravated for the past 1 week. He also complained of excessive worries, tension, and having sleep disturbance. There was no history of fever, weight loss, skin rash, loss of appetite, or backache. There was no past history of diabetes, hypertension, tuberculosis, polyarthritis, or any other significant illness. His family history was not contributory. Investigations revealed normal hemogram and urine examination. His blood urea, creatinine, LFT, electrolytes, and sugar levels were normal. Abdominal ultrasound revealed bilateral renal calculi with gross pelvicalyceal dilatation of right kidney and right hydroureter. Renal pyelogram showed non-functioning right kidney with multiple left-sided renal calculi. Tc99m diethylene triamine pentaacetic acid (DTPA) renogram revealed normal functioning and draining left kidney with a GFR of 55 ml/min and poorly functioning right kidney with a GFR of 10 ml/min. The patient underwent ureteroscopy with intra-corporeal lithotripsy of right kidney and bilateral DJ stenting. In addition, he underwent multiple sittings of extracorporeal shock wave lithotripsy. He had normal ESR, negative CRP, negative rheumatoid factor, serum ionised calcium 4.4 mg/dL, and serum alkaline phosphatase 86 IU/ml. Whole-body bone scan done revealed extremely hot focus involving the right humerus (head and proximal shaft) and left hemipelvis-features, suggestive of Paget's disease [Figure 1]. X-rays of right humerus and pelvis did not reveal any abnormality [Figure 2]. Magnetic resonance imaging (MRI) showed thickened cortex with altered signal intensity involving the right humerus head and proximal shaft and left iliac bone, commensurate with the bone scan findings [Figures 3 and 4]. A bone biopsy from left iliac crest was

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done, which showed irregular bony trabeculae lined by plump osteoblasts, with multinucleate osteoclasts and increase in cement lines [Figure 5]. Serum calcium was 9 mg/dL and PTH level was 12 pg/ml, both normal. Hence, a diagnosis of Paget's disease was made and the patient was started on biphosphonate therapy. He is under follow-up and has shown marked improvement in symptoms.

DISCUSSION

Paget's disease of bone is a localized bone remodeling disorder of uncertain etiology.[5] The onset of the disease is insidious and generally occurs later in life. Both men and women are affected, with a slight male preponderance. Bone that is remodeled by this pathological process becomes enlarged and mechanically weakened. Pain, skeletal deformity, and fracture may develop because of structurally inferior bone. Bone expansion may also cause various neurological compression syndromes, but most patients with Paget's disease are discovered because of an increased alkaline phosphatase level or an incidental X-ray finding and are asymptomatic.[2] In our case, the disease was detected during evaluation for nephrolithiasis and polyarthralgia. The patient had normal X-rays and normal serum alkaline phosphatase levels. Bone scan was done for polyarthralgia that showed the typical pattern of Paget's later confirmed by bone biopsy. Apart from the incidental, skeletal, or neurological presentations. Paget's disease is known to be associated with aortic valve, endocardial, and arterial calcification.[6] Nephrolithiasis and Peyronie's disease have also been associated with it.[4] Our case had severe bilateral nephrolithiasis, which was the primary complaint. The patient was also found to have depression and was started on treatment for the same. An increased incidence of depression has been reported in literature.[1]

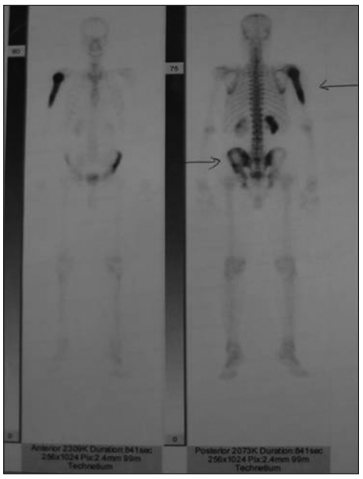

Figure 1: Bone scan done using Tc-99M labeled MDP-showing extremely high osteoblastic activity in the right humerus and left pelvic bone (as marked)

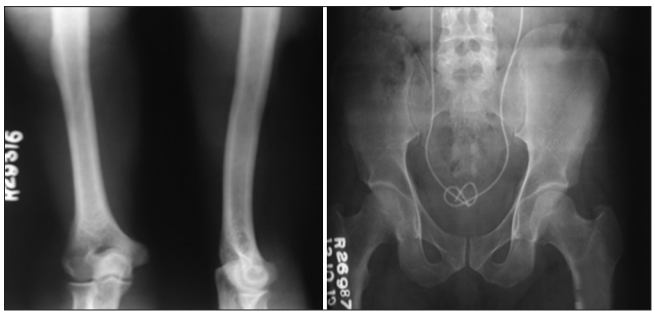

Figure 2: X-rays of right arm and pelvis showing no abnormality in bones

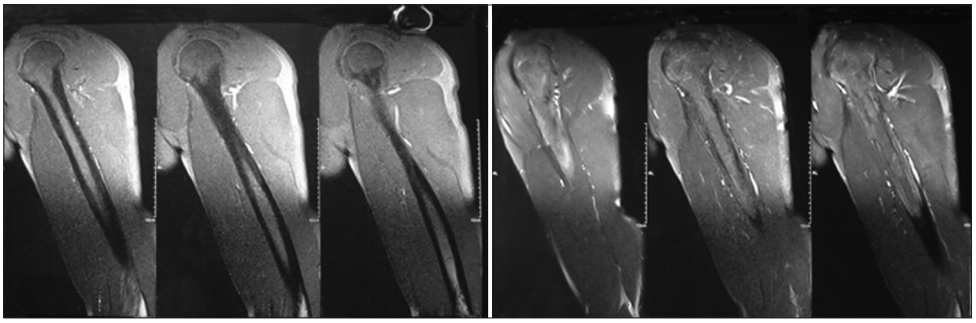

Figure 3: MRI right arm showing thickened cortex with altered signal intensity involving the right humerus head and proximal shaft

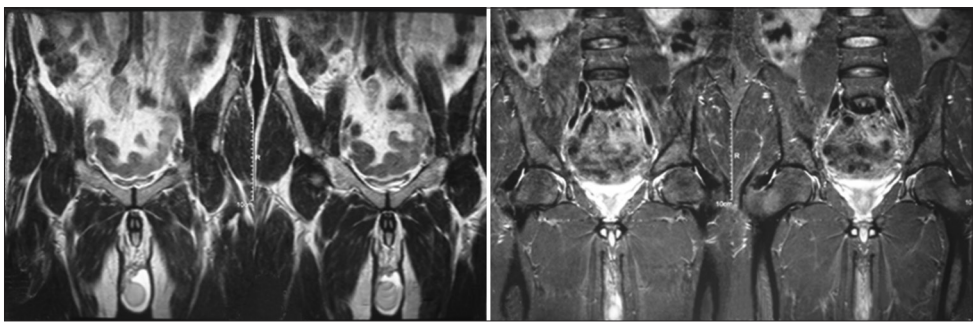

Figure 4: MRI pelvis showing thickened cortex with altered signal intensity involving left iliac bone

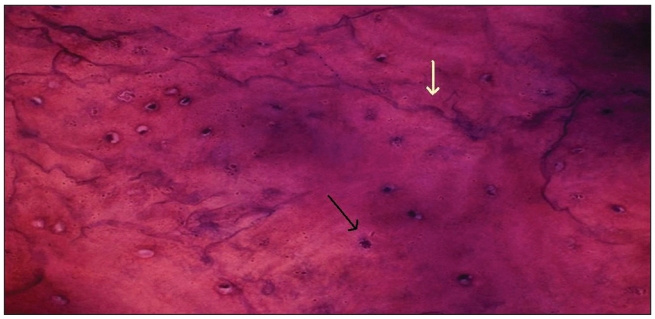

Figure 5: Bone biopsy from left iliac crest shows plump osteoblasts (black arrow) and increase in cement lines (white arrow)

Skeletal scintigraphy has had a major impact in diagnosis of Paget's disease because of the high sensitivity of the technique for detection of both increased vascularity and increased osteoblastic activity characteristic of the disorder. Radiography has much inferior sensitivity and bony Pagetic changes may be missed by it, as in our case. This has been reported previously as well.[7-9] Lesions become visible on X-rays when more than 30-50% of the bone has been resorbed. In fact, Smith *et al.*, in his study found that 73% of patients with positive bone scan and normal radiographs were symptomatic.[10] The scintigraphic appearances are fairly characteristic, showing intense accumulation of radiopharmaceutical throughout the affected part with uniform distribution. Pagetic lesions in long bones appear at the articular margin, progressing along the shaft, and producing a sharp V-shaped advancing edge like a flame, as was seen in our case in the humerus.

However, scintigraphy is not totally specific, as the differential diagnosis can be metastasis, metabolic bone disease, and even fibrous dysplasia if detected in later age group. Preservation or enhancement of normal bony anatomy is typical and differentiates Paget's from other scan abnormalities. Also, there is a clear distinction between normal and abnormal bone differentiating it from metabolic bone disease. In fibrous dysplasia, there is a lack of preservation of bony outline unlike Paget's. Metastatic prostate cancer and rarely breast cancer can mimic Paget's disease.[11] The pelvis and the spine are most problematic, particularly when there is a vertebral collapse or degenerative change. These considerations make radiological correlation advisable and even biopsy is occasionally necessary.

Clinical manifestations of Paget's disease in which pharmacologic therapy is recommended are symptoms resulting from active bone lesions like bone pain, headache with skull involvement, back pain due to pagetic radiculopathy or arthropathy, other neurologic syndromes, and fissure fractures. Symptomatic disease is the most common indication for pharmacotherapy.

Follow-up of these cases and monitoring the treatment response can be done by serum levels of biochemical markers like alkaline phosphatase. Patients with mono-ostotic disease and limited polyostotic disease may have normal levels of these markers. In such patients, scintigraphy is advocated to monitor disease status and treatment response.[12] In our case, despite polyostotic form, the biochemical markers were normal and hence bone scan will be utilized to follow-up the patient. In fact, it has been suggested that scintigraphy may better assess long-term response, since scintigraphic activity may persist after biochemical normalization.[13]

CONCLUSION

This case highlights the role of bone scintigraphy in detecting an unusual presentation of paget's disease in the form of nephrolithiasis, polyarthralgia, normal radiographs, and normal biochemical markers despite being in the polyostotic form.

REFERENCES

1. Reid IR, Miller P, Lyles K, Fraser W, Brown JP, Saidi Y, *et al.* Comparison of a single infusion of zoledronic acid with risedronate for Paget's disease. N Engl J Med 2005;353:898-908.
2. Altman RD. Musculoskeletal manifestations of Paget's disease of bone. Arthritis Rheum 1980;23:1121-7.
3. Eastell R. Biochemical markers of bone turnover in Paget's disease of bone. Bone 1999;24(5 Suppl):49-50S.
4. Whyte MP. Clinical practice. Paget's disease of bone. N Engl J Med 2006;355:593-600.
5. Siris ES, Roodman GD. Paget's disease of bone. In: Favus MJ, editor. Primer on the Metabolic bone diseases and disorders of mineral metabolism, 6th ed. Washington, DC: American Society for Bone and Mineral Research; 2006. p. 320-30.
6. Laroche M, Delmotte A. Increased arterial calcification in Paget's disease of bone. Calcif Tissue Int 2005;77:129-33.
7. Mazieres B, Jung-Rosenfarb M, Bouteiller G, Fournie A, Arlet J. Bone scintigraphy in Paget's disease. Rev Rhum Mal Osteoartic 1978;45:311-6.
8. Vellenga CJ, Pauwels EK, Bijvoet OL, Frijlink WB, Mulder JD, Hermans J. Untreated Paget disease of bone studied by scintigraphy. Radiology 1984;153:799-805.
9. Fogelman I, Carr D. A comparison of bone scanning and radiology in the assessment of patients with symptomatic Paget's disease. Eur J Nucl Med 1980;5:417-21.
10. Smith SE, Murphey MD, Motamedi K, Mulligan ME, Resnik CS, Gannon FH. From the archives of the AFIP. Radiologic spectrum of Paget disease of bone and its complications with pathological correlation. Radiographics 2002;22:1191-216.
11. Fig LM, Gross MD. Metastatic prostate carcinoma mimicking Paget's disease on bone imaging. Clin Nucl Med 1989;14:777-8.
12. Mari C, Catafau A, Carrio I. Bone scintigraphy and metabolic disorders. Q J Nucl Med 1999;43:259-67.
13. Ryan PJ, Fogelman I. Bone scintigraphy in metabolic bone disease. Semin Nucl Med 1997;27:291-305.

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