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Images

Mickey Mouse sign in a case of polyostotic Paget's disease

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ABSTRACT

Paget's disease of bone is an osseous dysplasia affecting the middle-aged and elderly population. It is most common in Northern Europe and Australia and is rare in Asia and Africa. We report a case of 50 year old menopausal female, who presented to us with features of polyostotic Paget's disease of bone. The classical radiographic and bone scintigraphic findings of the patient have been described.

Key words: *Alkaline phosphatase, Bone scan, Mickey Mouse sign, Paget's disease of bone*

Paget's disease, also known as osteitis deformans is a localized disorder of bone remodeling characterized by excessive bone resorption followed by an increase in bone formation. The exaggerated unchecked bone turnover gives rise to structurally disorganized woven bone, which is prone to pathological fractures. The clinical findings depend on which bones are affected and may range from an incidental radiological or biochemical abnormality to devastating musculoskeletal disabilities. Plain radiographs and bone scintigraphy may help in initial diagnosis and during follow-up to assess treatment response.

A 50-year-old menopausal female presented to our outpatient department with complaints of generalized bony pains and difficulty in getting up from squatting position for past 2-3 years. She also complained of painful restriction of motion at both hips, worse on right side. There was no history of any early morning worsening of symptoms or any history suggestive of small or large joint arthritis. Her symptoms had progressed gradually to an extent that she had difficulty in ambulation at the time of presentation. She denied any history of low trauma fractures or height loss. She had been reviewed by multiple physicians elsewhere and prescribed multiple courses of calcium and Vitamin D without any symptomatic relief. On examination, there was a severe restriction of both active and passive motion at bilateral hip joints. There was no evident bony deformity or spine deformity or areas of tenderness.

Investigative workup including blood counts, urinalysis, fasting plasma glucose, liver, and renal functions was unremarkable. Other investigation included normal serum total calcium (8.5; N 8.5-10.4 mg/dl), normal inorganic phosphorus (3.8; N 2.5-4.5 mg/dl), normal serum albumin (4.2; N>3.5 g/dl), and elevated alkaline phosphatase (ALP) (1586; 240-840 IU/L). Hormonal investigations included normal T4 (8.6; N 5.1-14.1 mcg/dl), thyroid stimulating hormone (3.4; N 0.27-4.2 uIU/ml), 8 am serum cortisol (12.2; N 6.2-19.4 mcg/dl), intact parathyroid hormone (46.04; N 15-65 pg/ml), and 25(OH) D3 (16; N > 20 ng/ml). BMD T score was −2.6 at spine and 3.7 at total hip.

Based on the clinical, biochemical and radiological workup, she was diagnosed as a case of polyostotic Paget's disease. Considering active disease status, she was planned for initiation of treatment with bisphosphonate after correcting vitamin D insufficiency. The patient was however lost to follow-up, and no definitive treatment could be initiated.

Paget disease of bone, first described in 1877 by Sir James Paget is an osseous dysplasia affecting the middle-aged and elderly population. The disorder is characterized initially by an increase in bone resorption, followed by a disorganized and excessive formation of bone, leading to pain, fractures, and deformities. The disease may, however, be totally asymptomatic, detected based on an elevated ALP alone. There can be monotonic or polyostotic forms and most commonly affected sites include pelvis, spine, skull, and long bones [1-3].

Classical radiological findings including cortical thickening, accentuation of bone trabeculae and increased bone size. Findings are frequently asymmetrical, manifesting as cortical thickening and cotton wool spots in skull, picture frame or ivory vertebrae in spine, coarse trabeculae, thickened iliopectineal line and protrusio acetabuli with degenerative joint disease in pelvis and cortical thickening with bowing deformities of long bones. Whole body bony scintigraphy with Technetium-99 m methylene bisphosphonate helps in quantifying the disease burden at the initial visit and may be useful in follow-up as well. It reveals increase uptake at the sites of disease activity. Increased scintigraphic uptake in the body and spine of the vertebrae assumes a peculiar shape and has been famously described as Mickey Mouse sign in Paget's disease [2] (Figs. 1-3).

Bone pain is the most common indication for initiating treatment in Paget's disease. Treatment options include calcitonin

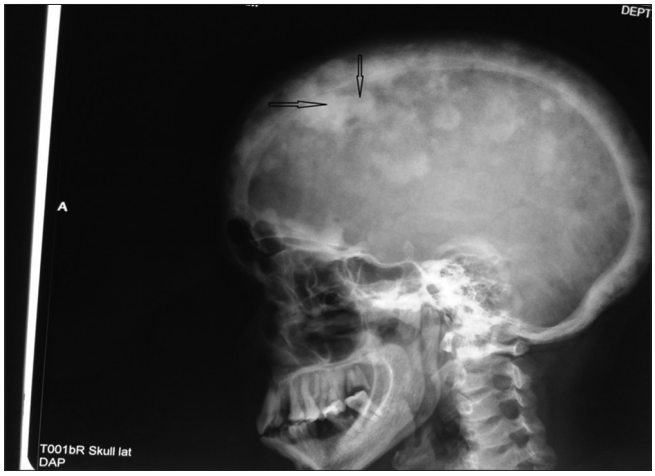

Figure 1: Plain X-rays skull showing thickened calvarium with mixed sclerotic and lytic lesion imparting cotton wool appearance to the skull (arrow)

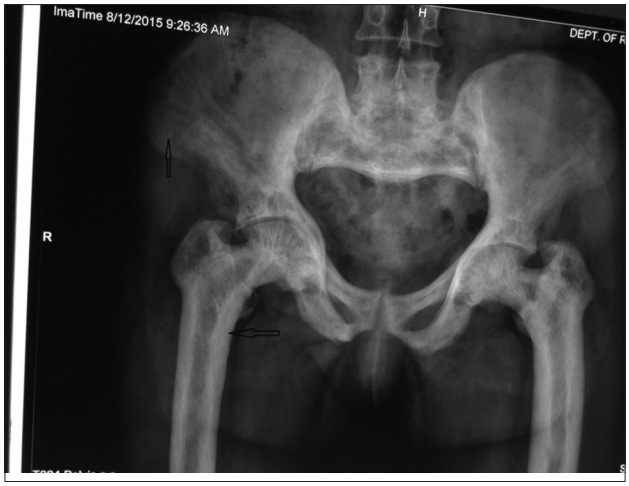

Figure 2: Plain X-rays pelvis showing accentuated coarse trabeculae (arrow) and cortical thickening (arrow), more prominent on the right side

and bisphosphonates with surgical treatment required in rare cases with significant bony pain and deformities [4-5].

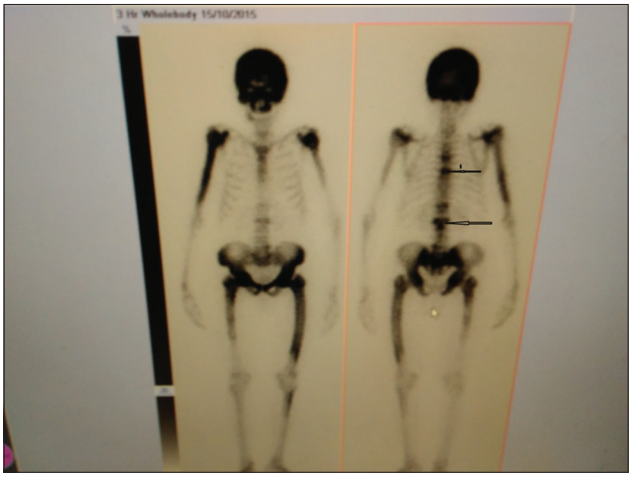

Figure 3: Technetium 99 m-methyl bisphosphonate bone scintigraphy showing intense radiotracer uptake at skull, bilateral humerus, pelvis, bilateral femur, and spine (Mickey Mouse sign marked by arrow)

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