

FEMALE WITH CLASSICAL PRESENTATION OF PAGET'S DISEASE : A CASE REPORT

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ABSTRACT

Paget's disease is a osteometabolic disorder of bone remodelling which is rare in India and Asian countries . Its diagnosis in asymptomatic patients is done by incidental radiographs and bone markers . Present case report in female with low backpain emphasizes the importance of including paget's as differential diagnosis of unusual backpain patients. Treatment with Bisphosphonates and follow up with serum markers can prevent complications and achieve adequate disease control.

KEYWORDS : Paget's Disease, Bisphosphonates, Low Backpain, Bone Markers, Metabolic Bone Disease.

Paget's disease (Osteitis Deformans) is an idiopathic progressive osteometabolic disorder of bone remodelling (Griz et al., 2006). It is characterized by involvement of a single or multiple bones with excessive osteoclastic resorption followed by increased osteoblastic activities(Cundy and Bolland, 2008). Paget's Disease is rare in India & Asian Countries (Hadjipavlou et al., 2002; Chakrabarty,1963; Sridhar, 1994). Men are affected more commonly than Women (Bhatt et al., 2006). It is common in Past Middle Age of > 50yrs. We report a case of female presented with low-back pain where paget's disease was the etiology.

CASE REPORT

We report a case of a 50 yr female with Low back pain and Right Thigh Pain of 9 month duration who had taken multiple consultations. Pain was non radiating and rest local Clinical Examination was normal. Lumbosacral Radiographs showed Dense Trabeculation forming Picture Frame like Configuration in L2 Vertebrae .(Figure 1). Suspicion of Paget's Disease was raised. Radiographs of Pelvis with both hips showed thickened trabeculae & Cortices (Figure 2). Skull X-ray showed Classical Radiographic Pattern (Cotton Wool Skull) of Paget's Disease (Figure 3). Lab Assesment revealed increased Serum Alkaline phosphatase Levels (760 IU/L) with Normal Serum Calcium & Phosphorus Levels. Tc99m Bone Scan was advised but couldn't be done because of cost constraints. Patient was treated with oral bisphosphonates

(Risedronate 70 mg weekly) for 6 months. After Treatment Patients Symptoms improved with near normal Serum Alkaline Phosphatase level.

DISCUSSION

Paget's Disease is common in Western Europe, Americas, and Australia and rare in Asian Population (Hadjipavlou et al., 2002; Chakrabarty, 1963; Sridhar, 1994). Majority (\approx 95%) of patients are asymptomatic (Papapoulos,1997) and detected incidentally due to Raised alkaline phosphatase or radiographic picture however in our case patient was symptomatic for last 9 months. Symptomatic patients may present with Pain, Deformity, Fracture and malignancy (<1%).Majority are polyostotic involving pelvis, thoraco-lumbar spine, femur and skull (Papapoulos,1997).

Paget's Disease is diagnosed primarily by characteristic radiological appearance of mixed lytic and sclerotic areas, thickened trabecula, bone expansio, cortical thickening, and deformity (Shankar et al., 2013). (Figure). Initially, X rays may be normal but bone scan is more sensitive and helpful in detecting polyostotic disease(Papapoulos,1997). Markers of Bone resorption (Urinary Pyridinium collagen Crosslinks) and Bone formation (Alkaline Phosphatase) are raised (Rosen, 1996). These markers are used for monitoring of disease activity & Treatment Guide. Bone Biopsy usually reveals a characterstic "Mosaic" Pattern with widened lamellae,

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Figure 1



Figure 2

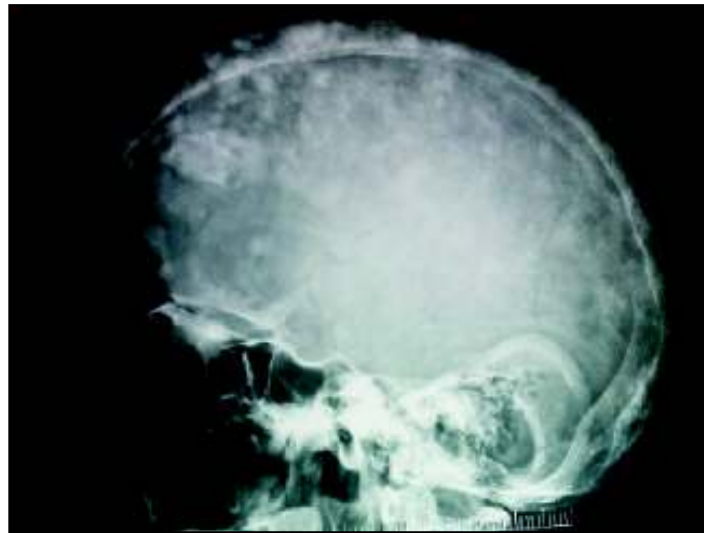


Figure 3

irregular cement lines and fibrovascular connective tissue.

In 1960, parenteral calcitonin was first used but because of its short half-life, frequent relapses after stoppage of therapy and resistance to treatment, its use is limited nowadays (Bhatt et al., 2006). Bisphosphonates (Oral and parenteral) are now the drugs of choice (Papapoulos, 1997). Pain relief, reduction in urinary

pyridinoline and deoxypyridinoline cross-links and reduction/normalization of alkaline phosphatase occur with adequate therapy. Treatment can be stopped after adequate control and the patient is followed closely every 3-6 months for bone markers (Alvarez, 2000). Treatment need to be restarted after 25% increase in the serum markers as compared with lowest achieved previous levels (Alvarez,

2000). Even after complete improvement quantitatively bone scan may show residual uptake in 20% cases (Bhatt et al., 2006).

CONCLUSION

Though paget's Disease is rare in Indians, but it should be considered in differentials whenever evaluating a low-backache patient in age group >50yrs. It is diagnosed by its classical radiographic features and raised serum markers. Bone scan, bone alkaline phosphatase and urinary pyridinoline/de-oxy pyridinoline are useful guide to therapy. Early suspicion and diagnosis can be helpful to prevent potential complications like pathological fractures, arthritis, hearing loss, high cardiac output heart failure (polyostotic) and rarely osteosarcoma. Currently, Second generation N-containing Bisphosphonates leads to normal or near normal bone turnover indices in a majority of patients.

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