Ivory Hemipelvis: A Presentation Of Paget's Bone Disease

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INTRODUCTION:

Paget's disease of the bone is a metabolic disorder involving atypical osteoclasts leading to abnormal bone remodelling. This disease is more common among the Western populations with a prevalence rate of between 0.3 – 7%, but report amongst the Asian populations is scarce. This case report aims to create awareness among the orthopaedic fraternity to a disease that is largely elusive in our region.

MATERIALS & METHODS:

A 68-year-old lady presented to us with 2 year history of left gluteal and hip pain which became worse 1 month prior to consultation. The pain is localized to the left gluteal and hip, on and off in nature, and is associated with nocturnal pain. The pain had caused limitation in walking distance to 10 metres and she was unable to perform her prayers in the usual manner. The pain Visual Analogue Score was 8 out of 10 and relieved by NSAIDs. She also gave a significant history of gradual loss of appetite and a weight loss of 5 kg over the past 1 year. She denied other joint pain, back pain, leg claudication or any history of trauma prior. There was no altered bowel habit and no previous infective contact. She has no significant family history of malignancy.

RESULTS:

Her blood investigation showed a Hb of 10.4 g/dL, WBC of 13.7x 10^9/L, elevated ALP at 470U/L, c-reactive protein of 200mg/L and normal serum calcium level. Pelvic radiograph showed diffuse sclerotic changes of the left hemipelvis with cortical thickening (Figure 1). MRI of the pelvis showed extensive marrow signal abnormalities of the left hemipelvis with mild expansion (Figures 2). HPE showed thickened and disorganized bony trabecular pattern. There were cement lines seen along the coarsened and enlarged trabeculae. In many areas, the marrow spaces were calcified with some area demonstrating replacement of marrow space with fibrous tissue. There were no obvious osteoblastic or osteoclastic cells seen. No granuloma or atypical cells suggestive of malignancy found. These findings are consistent with the diagnosis of Paget disease of the left hemipelvis.

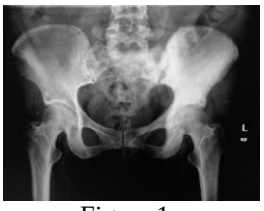

Figure 1.

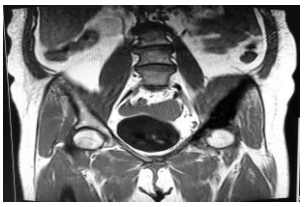

Figure 2.

Figure 1: Pelvis AP radiograph.
Figure 2: T1-weighted coronal MRI of pelvis

DISCUSSIONS:

Paget's disease of the bone is a condition usually diagnosed in the fifth decade of life. Most pagetic patients complain of bone pain or fracture following a trivial injury. Others may be picked up incidentally on radiographic findings. The commonly affected bones are the pelvis (70%), femur (55%), lumbar spine (53%), skull (42%), and tibia (32%). Indolent infection with paramyxovirus and respiratory syncytial virus has been implicated as possible cause for Paget's disease. The distinctive radiographic features and isolated serum ALP elevation are diagnostic of this disease and hence biopsy is rarely needed.

CONCLUSION:

The diagnosis of Paget's disease should be entertained in patients with raised ALP and osteoblastic lesion in the hemipelvis.

REFERENCES:

Merashli M et al., Paget's Disease of Bone among Various Ethnic Groups. Sultan Qaboos Univ Med J 2015 Feb; 15(1):e22-e26.