

Clinical Characteristics of Paget Disease of Bone from Turkey

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OBJECTIVES

Prevalence of Paget's disease (PD) of bone shows geographical difference. While highest prevalence reported from western countries it is rare in eastern countries. Clinical and demographical presentation of the disease may differ between the populations.

Aim: In this study we evaluate clinical and demographical parameters of Paget's disease patients followed from endocrinology clinics in Turkey.

METHODS

Method: An invitation was sent to all tertiary endocrinology clinics in Turkey to complete a survey on demographic clinical, laboratory parameters as well as used treatment modalities of patients with Paget's Disease. Sixteen centers from 12 cities reported 185 PD cases clinically or histologically proven.

RESULTS

A cohort of Paget's disease from Turkey has female preponderance (FM/M :105/80) with a mean age of 57 ± 10 years at onset. Its clinical features are bone pain, back pain and headache Fracture and typical skeletal involvement reported in 5 and 18 patients respectively. Only 2 patients have family history for PD. %67.5 (n:125) patients have polyostotic disease. Skull (41.6%), pelvis (53.5%), spine (41%) and femur (25.4%) are being the commonly affected bone sites. Seventeen patients with skull involvement reported to have hearing loss.

The biochemical profile at diagnosis had a mean alkaline phosphatase (ALP) 5520 ± 652 IU/L (Range 280 to 5762 IU/L), serum calcium: 9.2 ± 1 mg/dl, iPTH : 73.9 ± 65 pg/ml, 25OHvitD : $32,6 \pm 27$ ng/ml. Five patients wasn't need treatment.

Intravenous bisphosphonates was the most commonly used drugs (42% and 23% of patines treated with zoledronic acid 5mg and pamidronate 60 -90 mg respectively) remaining given high dose oral bisphosphonate (alendronate 40 mg/day or risedronate) Most cases respond well with decreased Serum ALP level (117 ± 114 IU/L) at the 6th month of the therapy. Ten patients reported partial remission and 5 patient relapsed according to ALP levels at the first year of the treatment. Duration of follow up was 7.5 ± 6.5 yrs.

CONCLUSIONS

Conclusion: In this group of PD from Turkey we observed female predominance. Pain and Bone Involvement were the predominant clinical features no significant family history. Polyostotic disease with pelvis, skull, spine and femur being the commonly affected bone sites. Classical clinical and biochemical features responds well to intravenous or high dose bisphosphonate therapy and need a lifetime follow-up