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Abstract

Pagets disease of bone is a rare condition in India and other Asian countries, although it is a common metabolic bone disorder in west. The disease affects elderly and exact etiology is not known. It clinically presents as longstanding bone pain and deformity. We report a case of pagets disease in India affecting humerus and presented with pathological fracture twice before being diagnosed as pagets disease. Fracture was treated conservatively and patient was started on alendronate. Patient responded favourably at the end of six months of treatment.

Introduction

Pagets disease (PD), described by Sir James Paget in 1877, is a metabolic disorder affecting bone remodeling. It is a chronic disorder resulting in bone thickening and deformity due to abnormally increased bone resorption and formation. It is commonly seen in the elderly population. It mostly occurs in western population with overall prevalence upto 2%. Britain has the highest prevalence of 4.6% and in Europe it is upto 2% [1]. Until recently, PD was considered almost a non existent entity in asian and African populations, with only few cases reported from this region, but this perception has changed now and recent studies have reported that PD does occur in the Indian and asian population although not commonly [11]. The exact cause of rarity in this region is not known, but low index of suspicion and underreporting could have resulted in lower number of cases in Indian literature. It can occur in both polyostotic or monoostic forms. Commonest form is polyostotic and the usual sites involved are pelvis, lumbar spine, femur [3]. Clinical presentation depends on the site involved such as bone pain, backache, joint pain, headache, fracture [4]. We present here a case of pagets disease of humerus presenting with recurrent fractures.

Case Report(s)

A 65 year old male presented with severe dull aching pain of left arm since eight weeks. Patient had history of trivial fall four weeks back and had suffered a fracture at lower third of the humerus. He was treated conservatively with a POP cast at a local hospital. Now, patient had a continous dull aching pain over the whole arm. Patient also had a similar episode of fracture of the humerus one year back after a fall from his bed. The pain before the episode of fall was intermittent and was relieved on NSAIDS. There was no history of any constitutional symptoms like fever, loss of weight. Pain was localized to the arm and non radiating, and increased on excessive physical activity. On examination, there was local tenderness of the whole of humerus. There was no distal neurovascular deficit. Radiographs revealed the whole of humerus showing abnormal bony architecture in the form of lytic and sclerotic lesions with thickened cortex with evidence of healing fracture at the lower third of humerus and healed fracture at the upper half. All blood investigations were normal except that serum alkaline phosphatase was raised upto 780 IU/L. Whole body dual intensity technicium bonescan was performed and the whole of left humerus showed increased radioactive uptake with some uptake also at the right ilium. A biopsy of the affected bone showed irregular prominent cement lines with thickened trabeculae suggestive of pagets disease. Patient was put on NSAIDS and bisphosphonates. Alendronate was started at dose of 40mg/day for six months. Pain improved significantly during course of treatment and alkaline phosphatase levels reduced to 326 IU/L at the end of six weeks. The fracture healed completely by the end of 8 weeks. Patient has no complaints at the end of six months of treatment.

Discussion

Pagets disease(PD), since its first description by Sir James Paget, has been studied extensively and a lot of views about this disease are changing as regarding its peculiar geographical distribution, etiology, and treatment. PD has always been regarded as a disease affecting european population especially great Britain with prevalence upto 4.6%. Australia and North America also have a high prevalence [1]. The disease has been considered rare in Asian and African population. In India, according to recent studies the total number of reported cases are only about 100
These reports along with studies from China and Japan have shown that PD is not totally absent among Asians [6,12]. Studies have also shown changing epidemiological patterns such as declining prevalence in regions like New Zealand and Great Britain, increase in age of presentation, reduction in severity [2,8]. Age of onset is usually above 55 years but familial cases may be of younger age [7].

Etiology of the disease is still undergoing extensive research. Possibility of viral etiology due to presence of inclusion bodies has been proposed. Paramyxovirus, respiratory syncitial virus and canine distemper virus have been studied as a cause, but conclusive evidence is not established. Environmental influences and familial inheritance have been proposed because of the geographical pattern of distribution, although no environmental cause is established but studies have shown presence of germline mutations affecting SQSTM1/p62 gene in both familial and sporadic cases of PD [10].

The disease is characterized by abnormal remodeling of the bone and is divided into three phases. Initially there is osteolytic phase in which excessive bone resorption occurs due to overactivated osteoclasts, then mixed phase with features of resorption and sclerosis both. The last phase is the burnt out phase or sclerotic phase where new bone formation occurs irregularly and excessively by osteoblasts. PD can occur in either polyostotic or monostotic forms. Polyostotic form is the most common type but, recent trends show that number of monostotic cases are increasing up to 40% [8,9]. It also accounts for another changing trend of reduction in severity of the disease due to increased proportion of single bone involvement. Clinically the most common presentation is bone pain seen in more than 90% cases. Other presenting features are fractures, deformities or neurological complications. Very often, the disease is asymptomatic and is detected as an incidental finding, usually in monostotic forms.

Biochemical markers such as raised serum alkaline phosphatase has high sensitivity. Bone resorption markers like type 1 procollagen peptide have also been used but are not as sensitive. Radiographs and wholebody bone scan are the most important radiological investigations in the diagnosis of PD. Radiographs show features depending on the stage ranging from lytic lesions, thinning of cortex and sclerotic features like trabecular accentuation, thickened cortex. The commonest features seen are of sclerotic phase [4]. Wholebody bone scintiscan is considered more sensitive than radiographs in pagetic lesions but is less specific and has to be differentiated from other bone tumours and metastases. Bonescan findings substantiated by radiography help in establishing a diagnosis. Bone biopsy is indicated to differentiate from tumours and other disorders of bone remodeling [8]. Biopsy in PD shows irregular trabeculae with cement lines and increased number of osteoclasts and osteoblasts. Biopsy is also indicated in complications such as osteosarcoma.

Pagets disease is known to cause complications the manifestations depend on the site. Worsening pain, deformity, and fractures are usual complications. Fractures occur in about 10% of cases involving long bones [3]. Neurological complications when skull is involved are loss of hearing, headache etc. Joint related complication such as degenerative arthritis in segments involving joints [5]. Malignancy occurs in less than 1% of cases with osteosarcoma being commonest followed by fibrosarcoma and chondrosarcoma. Long term follow up is needed to detect such untoward complications.

Treatment of pagets disease is mainly medical. Bisphosphonates are the drug of choice [12]. Introduction of bisphosphonates has vastly improved the results with all of the patients and even complications except sarcoma and deafness responding to it [13]. Compliance of the patients has also improved because of use of oral bisphosphonates. Alendronate has shown better results in comparison to etidronate [14]. Recommended dose of alendronate is 40mg/day and studies comparing with lower doses (20mg/day) have shown that decrease in serum alkaline phosphatase is lesser than levels obtained at higher doses [12]. However, Joshi et al have shown that lower doses may be sufficient in Indians and sap levels in most cases reached baseline by six months in their study [14].

**Conclusion**

In conclusion, pagets disease in India is not as rare as has been thought to be. Monostotic pagets disease may be asymptomatic for a long time and present when complications such as fracture occur. Pagets disease must be one of the differential diagnoses in such cases especially when serum alkaline phosphatase is raised. Alendronate is very effective in the management and also prevents complications. Long term follow up is must to watch for malignant transformation.

**References**
Illustrations

Illustration 1

whole body bonescan showing increased uptake at left humerus and ilium

Illustration 2

Radiograph showing healing fracture at lower third humerus and old healed fracture at upper half
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