Paget’s Disease of Bone in a 85 Year Old Woman
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Abstract
Paget’s disease of bone is a rare disorder in India. Patients are usually asymptomatic and incidental radiological investigation invariably clinches the diagnosis. Increased alkaline phosphatase, X-ray, CT scan and scintiscan of bone can be done to confirm the diagnosis. Here we report a case of an 85 yrs old lady who had bilateral hearing loss for last 10 yrs, was admitted incidentally for some other complaints and on investigation diagnosed to have advanced Paget’s disease of bone.

Introduction
It is now nearly 100 yrs that Sir James Paget described the condition he called “Osteitis Deformans”. The disease has an extraordinary geographical distribution. It is almost unknown in Africa, India and China.1 But Paget’s disease of bone (PDB) is now reported from different centers of India during the last few years.1,2,7,8 We present in this report an 85 yrs old Indian lady who was diagnosed to have PDB while being investigated for headache and drug induced vomiting.

Case report
An 85 yrs old Hindu female from Jajpur, Orissa was admitted to the hospital with chief complaints of headache and vomiting for 2 days. She had history of impaired hearing for last 10 yrs. She was not a known diabetic or hypertensive or on any chronic medication for any disease. On physical examination, her pulse was 70/min (regular), BP 140/90 mm Hg, no pallor, no oedema and JVP was not raised. She was conscious, oriented and afebrile. She had bilateral hearing impairment due to conductive deafness (BC>AC), bilateral visual loss due to cataract. Motor and sensory examination did not reveal any abnormality. Cardiovascular and respiratory examinations were also normal.

Her investigation showed Hb 8.8 gm%, TLC 8000, P.<sub>E</sub>B.<sub>L</sub> M, FBG 102 mg/dL, urea 31 mg/dL (normal 7-20 mg/dL), creatinine 1.1 mg/dL (normal 0.6-1.2 mg/dL), sodium 136 meq/L (normal 136-146 meq/L), potassium 4.1 meq/L (normal 3.5-5 meq/L), ESR 40 mm/h (normal 0-20 mm/h), serum total calcium 8.9 mg/dL (normal 8.7-10.2 mg/dL), alkaline phosphatase 750 units/L (normal 41-133 units/L). As the patient presented with headache and vomiting in old age, CT scan of brain was done. It showed thickening of inner and outer table of skull, with prominent ventricles and cortical sulci suggestive of cerebral atrophy and diffuse fatty infiltration of the marrow (Figure 1). Bone Window and scanogram showed sclerosis of the base of the skull (Figure 2) and focal areas of patchy sclerosis involving parietal bones.

Fig. 1: CT scan of brain showing thickening of outer & inner table of skull with prominent ventricles and cortical sulci and diffuse fatty infiltration of the marrow

Fig. 2: CT scan of brain showing sclerosis of the base of the skull.
and widening of diploic space (Figure 3). X-ray of the skull (lateral view) showed widening of diploic space and calvarial thickening (Figure 4). X-rays of upper and lower extremities revealed osteoporotic changes.

Discussion

PDB causes malfunction in the normal process of bone remodeling. Normally bone is continuously breaking down and remodeling. This normal process of destruction and remodeling is somehow altered in PDB. When an area of bone is destroyed in PDB, the bone that replaces it is soft and porous. Many patients with PDB are asymptomatic and are often diagnosed by an elevated alkaline phosphatase level on routine blood investigation or from an abnormal skeletal radiograph done for other indications. Most common symptom is bone pain at the site of pagetic involvement. Patient may also present with bony deformity, pathological fracture, high cardiac output, or features of nerve compression. Progression of the disease is characterized by initial lytic phase, a mixed lytic and blastic phase and the sclerotic or burnt out phase seen late in the disease process. The radiological signs of these three phases can be found in the same patient in different locations.

The most common consequence of Paget’s involvement of skull is hearing loss which can be either conductive or sensorineural. Cause of conductive deafness is pagetic involvement of the bones in inner ear. Vertebral involvement can cause kyphosis and compression of the spinal cord. Skull involvement may also produce cranial nerve palsies, cerebellar dysfunction or obstructive hydrocephalus. Activity of the disease can be assessed by measurement of bone thermolabile alkaline phosphatase, and indices of bone resorption like urinary excretion of total hydroxyproline, pyridinoline and deoxypyridinoline. X-ray and scan picture of PDB shows destructive process affecting the outer table and sparing the inner table. Later on osteoblastic activity supervenes and seen as sclerotic areas on a background of lytic lesions. This is described as “cotton-wool appearance” in X-ray skull. Soft tissue extension causes typical “sunburst” picture. CT shows the extent of the disease with greater accuracy and is useful if reconstructive surgery is contemplated. MRI shows the yellow marrow is maintained and hyperintensity in T1 image. The areas of hypointensity in T1 indicate sclerosis. Osteoblastic metastasis from prostatic carcinoma and lymphoma are important differential diagnosis. Metastatic lytic areas are less well delineated than pagetic lytic areas and in pagetic bones cortical thickening and adjacent thickened trabeculae are characteristic. Skeletal scintigraphy is diagnostic of this metabolic disorder and whole body surveys are needed which often shows multiple sites of involvement with varying degree of activity.

We are reporting a case of PDB from Orissa. Peculiarity of the patient was not only her extremely high age of presentation (85 years) but also she was almost asymptomatic even at that age in spite of far advanced disease state. PDB is almost unknown in Africa, India and China. Recently PDB is reported from different centers in India. In a multicentric study from different parts of India, 21 cases of PDB were diagnosed between 1999 and 2005. No case was reported from eastern India. Another retrospective study from south India reported 51 cases. There are some other isolated case reports of PDB in recent years, one each from Chennai, Lucknow and Manipal. Our case is reported from Orissa where PDB is unknown.

In the multicentric study of 21 patients, mean age at diagnosis was 49.2±17.6 years; common presenting manifestations were bone pain, backache and headache. The most commonly involved bones were lumbar vertebra and skull followed by pelvis. Biochemical profile showed serum calcium 9.1±0.5 mg/dl and phosphate 3.9±0.53 mg/dl. Serum alkaline phosphatase was elevated in all but two patients. Mean alkaline phosphatase level was 1514±1168 IU/L and ranged between 550-6926 IU/L. In a retrospective study of 51 patients of PDB at a centre in southern India, the mean age of presentation was 56 years and biochemical analysis showed the mean serum alkaline phosphatase to be 690 U/L at the time of presentation. In the case reported from Manipal, serum alkaline phosphatase level was 2721 IU/L and serum calcium, phosphorus levels were normal. The report from Chennai was similar. In the case...
report from Lucknow, serum calcium, phosphorus and alkaline phosphatase were normal and diagnosis was confirmed by CT-guided bone biopsy. Our patient had serum calcium level of 8.9 mg/dl and high alkaline phosphatase level of 750 U/L. Thus very high alkaline phosphatase is the single most important biochemical investigation to suspect PDB. In the multicentric study of 21 patients, radiologic investigation showed sclerotic and lytic changes in all patients as well as bone deformity and osteoarthritis in eight patients. In the case report from Manipal, skull radiograph showed marked calvarial thickening and predominant sclerosis of inner table. Sclerosis of the base of the skull was noted around foramen magnum leading to platibasia and basilar invagination. CT scan showed “cotton wool” appearance of cranial vault due to combined osteoblastic and osteolytic activity. Dilated lateral and third ventricle with periventricular seepage was noted suggestive of obstructive hydrocephalus. The case report from Lucknow showed extensive sclerotic lesion in the right hemipelvis. In our patient, X-ray of the skull (lateral view) showed widening of diploic space and calvarial thickening. CT scan of brain showed thickening of inner and outer table of skull, with prominent ventricles and cortical sulci suggestive of cerebral atrophy and diffuse fatty infiltration of the marrow. Bone window and scanogram showed sclerosis of the base of the skull and focal areas of patchy sclerosis involving parietal bones and widening of diploic space. In the case report from northern India the coexistence of PDB with metastatic carcinoma of breast was documented. The other report from Manipal has depicted a case of PDB complicated by hydrocephalus and dementia.

Our patient had bilateral deafness, typical X-ray and CT scan findings e.g. thickening and separation of outer and inner table of skull bones, focal areas of patchy sclerosis and high alkaline phosphatase. Thus our case is reported because of its rarity and classical way of presentation.

References