Case Report

Paget's Disease of skull of presenting as Hydrocephalus

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ABSTRACT

Paget's disease is a focal bone disorder associated with increased bone turnover manifesting as abnormal bone integrity. It is characterized by abnormal bone modeling and remodeling due to increased osteoclastic activity followed by inadequate osteoblastic repair. Rarely, it can affect the skull causing hydrocephalus. It is common in western countries but rare in Asians.

Keywords : Paget's, osteoblastic, osteoclastic, remodeling.

Introduction :

Sir James Paget first described this disease in 1877. Also known as osteitis deformans, it is the second most common metabolic disorder of bone after osteoporosis[1]. Initially it begins with osteoclastic hyperactivity with bone resorption followed by increased osteoblastic activity. The end result is a structurally disorganized (woven), poorly mineralized and a weak bone prone to fracture. It most commonly affects older population (age > 50 years) and can be monostotic or polyostotic. Patients can be symptomatic or asymptomatic depending on the disease extent.

Paget's disease of bone (PDB) is rare in the Indian subcontinent[2]. There are very few cases reported with PDB as a cause of hydrocephalus. Herein, we report an elderly woman with hydrocephalus due to PDB.

Case Report :

A 65 year old woman presented to the hospital with complaints of multiple falls since 1 week. It was associated with difficulty in walking. She also expressed concerns about her memory problems and irritable mood. Her past medical history was significant for hypertension and a cerebrovascular episode. Patient was currently on Amlodipine 5 mg once daily. On examination, her pulse rate was 86 per minute, regular; blood pressure was 140/90 mm of Hg. Neurologic examination revealed gait with short steps and a tendency to fall. It was slow but normal for her age. Based on the history and symptoms we suspected Central Nervous system (CNS) pathology. Cerebral and cerebellar function was normal. Eye and ear examination were normal.

Serum alkaline phosphatase (ALP) was elevated at 462 IU/L (normal 33-96 IU/L). Complete blood count, serum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), serum calcium and serum phosphorus were with in normal limits. Computed tomography (CT) scan of brain revealed mild expansion of calvarium with mixed lytic and sclerotic areas ***(Fig. 1).*** There was moderate dilatation of third and both lateral ventricles with mild periventricular edema ***(Fig. 2).***

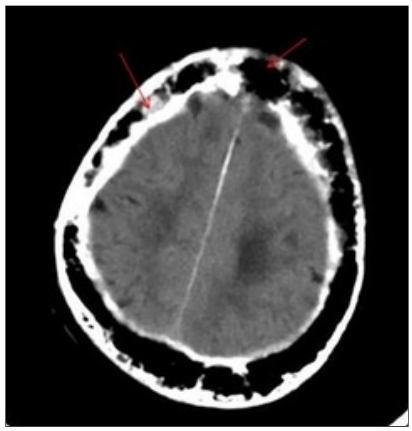

Figure 1 : **Axial CT image showing mixed lytic- sclerotic areas (red arrows) involving the bony calvarium of the skull**

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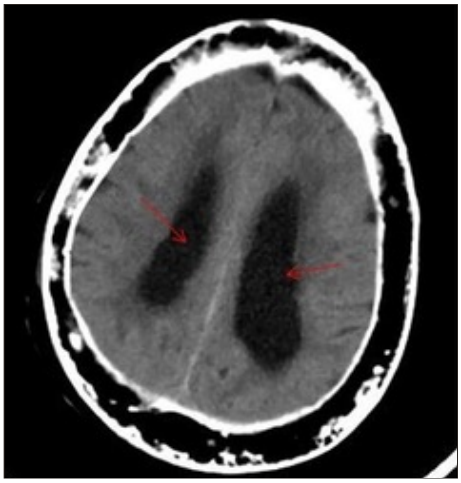

Figure 2 : **Axial CT image showing dilated ventricles (red arrows)**

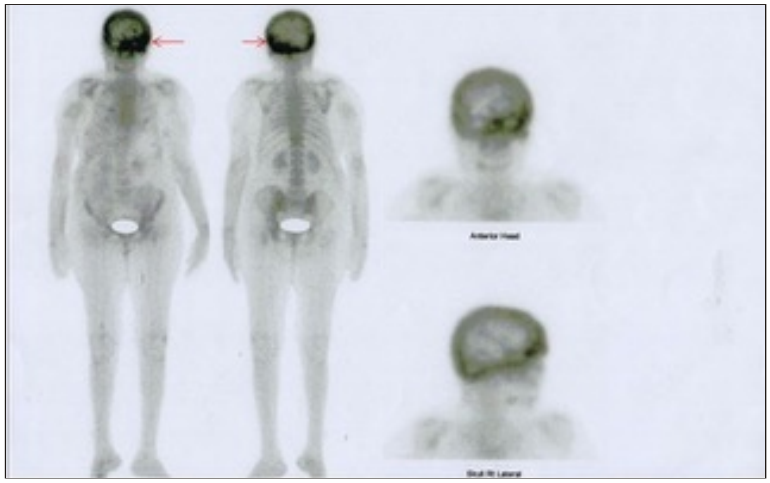

Figure 3 : **Nuclear bone scan showing diffuse osteoblasticover activity involving the skull (red arrows)**

Nuclear bone scan revealed diffuse osteoblastic overactivity of skull ***(Figure 3).*** Cerebrospinal fluid sample was collected after lumbar puncture and it revealed normal opening pressure and cell counts. Patient underwent a ventriculoperitoneal shunt procedure. CT scan of brain post procedure was done ***(Figure 4).*** Her symptoms improved significantly after the procedure.

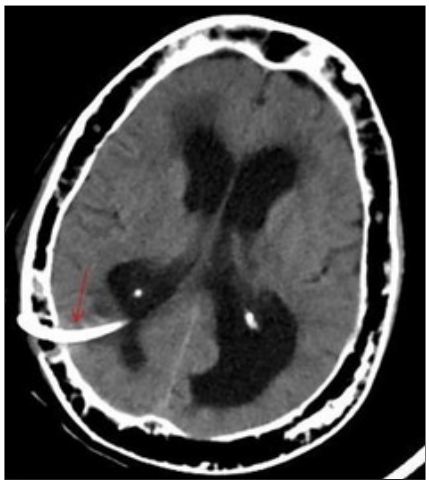

Figure 4 : **Axial CT image showing ventriculoperitoneal shunt (red arrow)**

Discussion :

Paget's disease is a chronic condition causing increased bone remodeling. This can lead to bone pain, deformity, pathological fracture and neurologic complications. It can affect any bone, but there is a predilection for pelvis, spine, femur, tibia and skull. It has an insidious, asymptomatic course and strong suspicion is needed for diagnosis. Risk factors include increasing age, male sex and white ethnicity. Pain is a very common symptom; it can be skeletal, neurologic, and joint or muscle origin[3].

Diagnosis is achieved by X-rays and radionuclide bone scan. In the earliest stage, osteolytic lesions can be seen in skull or long bones. It is followed by sclerotic phase characterized by mixed lytic and sclerotic areas. Most efficient modality is bone scan. CT skull and MRI brain can are done in cases of suspected hydrocephalus. ALP is the most useful marker for diagnosis and to evaluate response to treatment. The levels of ALP correlate with the disease extent. It is usually diagnosed with radiology while evaluating raised ALP levels unexplained by other causes. Hydrocephalus as a complication of Paget's disease is rare. Obstructive and non-obstructive hydrocephalus have been reported in the literature. Patients can present with ataxia, confusion, altered memory, features of Parkinson's disease and urinary incontinence.

Our patient had monostotic Paget's disease of skull. The most prominent feature was hydrocephalus. It occurs due to involvement of skull bone and blockage of aqueduct of sylvius. Other manifestations were gait disturbances and memory problems. There are few reports of Paget's disease

with normal pressure hydrocephalus. Goldhammer et al. reported a case of ataxia and dementia due to hydrocephalus related to Paget's disease of the skull[4]. Gottschalk described these findings in 1973. Paget's disease of the skull can lead to obstructive or non obstructive hydrocephalus.

Conclusion :

Paget's disease can present with neurological symptoms. If the neurological examination is non-significant, other conditions should be suspected. PDB can present with normal pressure hydrocephalus (NPH) and NPH could reveal an underlying Paget's disease of bone.

Consent :

Written informed consent was obtained from patient's legal guardian.

Conflicts of Interest :

The authors declare that there are no conflicts of interest.

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