

Case Report

Vertex extradural hematoma in association with Paget's disease of the skull

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Paget's disease of skull usually causes neurological complications such as basilar invagination, cranial neuropathies, etc. Occurrence of extradural hematoma in association with Paget's disease of skull is rare.

A 48 year old man presented with headache and right upper limb weakness, two days after a fall from scooter. CT Scan of brain showed a large extradural hematoma at the vertex with the cranial vault showing features of Paget's disease. At surgery, no skull fracture or injury to the superior sagittal sinus was evident. There was diffuse oozing from the inner table of the skull, which showed features of Paget's disease. The extradural hematoma was evacuated and the patient made good recovery. He deteriorated a few hours after surgery. Follow up CT Scan showed diffuse brain swelling with minimal recollection of hematoma. Re-exploration showed a small recollection which was evacuated and the part of the oozing diseased skull was excised. The patient recovered completely.

This is the first reported case of vertex extradural hematoma in association with the Paget's disease of skull. The unusual features are the absence of skull fracture and injury to the superior sagittal sinus. The increased vascularity of the skull due to Paget's disease has caused the hematoma by diffuse oozing from the inner table.

Key Words: Vertex extradural hematoma, Paget's disease of skull, extradural hematoma in Paget's disease

Introduction

Paget's disease, also known as osteitis deformans, is a disease of unknown etiology affecting about 3 percent of the population above the age of 40 years. It may be localized to one or more bony regions without any significant clinical findings; or it may be widespread and severe, producing extensive skeletal abnormality. The disease is characterized by excessive and

abnormal remodelling of bone, with bone resorption and formation in the active phase and diminished rate of bone turnover in the quiescent phase. These processes produce the characteristic pathological and radiological changes. Skull involvement in Paget's disease occurs in 25 to 65% of cases. The neurological disorders due to the Paget's disease of skull include basilar invagination, cranial neuropathies, hydrocephalus, etc.^[1] Intracranial extradural hematomas have not been commonly associated with the Paget's disease of skull. Literature search provided only 4 reported cases so far.^[2-5] Extradural hematomas occurring at the vertex have also been relatively rare. There is no previous report of the association of vertex extradural hematoma with the Paget's disease of skull.

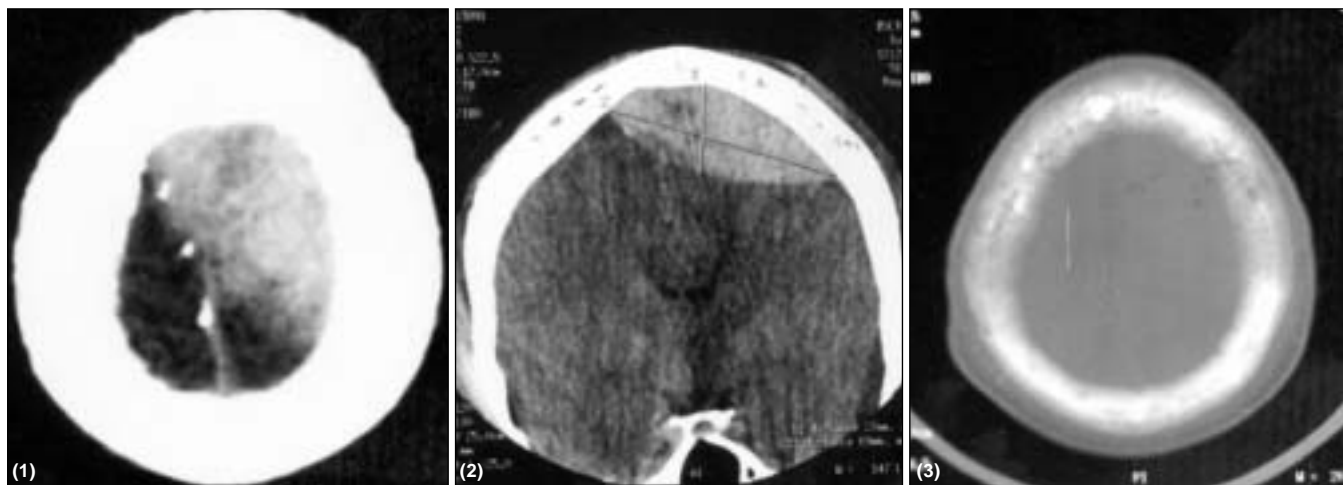
Case Report

A 48-year-old man was admitted to the hospital, 2 days after fall from scooter, with severe headache and right upper limb weakness. He had transient loss of consciousness at time of accident. At the time of admission, he was fully conscious and alert; he had normal optic fundus and grade 3 power in right upper limb. His vital parameters were normal. He had no external scalp injury. Computerized Tomographic Scan (CT Scan) of head showed moderate sized extradural hematoma at the vertex, more towards the left side, (Figure 1) well brought out in the coronal cuts. (Figure 2). The skull vault showed widened diploic space, with coarse trabecular pattern, suggestive of Paget's disease of skull. (Figure 3).

A frontoparietal vertex craniotomy extending across the midline was performed. There was no skull fracture. The skull was very soft, friable and very vascular, characteristic of Paget's disease. About 100 ml of extradural clot was evacuated. There was no injury to the superior sagittal sinus. There was diffuse ooze from the inner table, which was controlled with bonewax. The patient recovered well and his right upper limb weakness improved immediately after surgery. About 10 hours after surgery, he started deteriorating in conscious level, with a Glasgow Coma Score of 7. CT Scan at that time showed a small recollection of hematoma and diffuse brain swelling with ob-

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Figures 1-3: (1) Axial CT scan brain showing the vertex extradural hematoma. (2) Coronal CT scan brain showing the vertex extradural hematoma more clearly. (3) Bone window CT picture of skull showing widened diploe with coarse trabecular pattern suggestive of Paget's disease

literation of basal cisterns. Immediate re-exploration, evacuation of recollected hematoma and excision of the diffusely oozing bone flap were performed. The patient made good recovery and was discharged from the hospital after a week. Histopathological examination of the excised bone confirmed the diagnosis of Paget's disease.

Discussion

Extradural hematoma in association with Paget's disease has been rare. Drapkin has reported epidural hematoma occurring in an 82-year-old man with Paget's disease of skull.^[3] He has analyzed various pathogenic mechanisms operating in Paget's disease which may contribute to the development of extradural hematoma: (i) The soft nature of the affected skull may result in fracture after minimal trauma; (ii) Increased blood flow in the affected bone due to increase in the number of arterioles and capillaries, which may facilitate the development of epidural hematoma after trauma; (iii) Decreased dural adherence due to the changes in the physical quality of the affected skull; (iv) Atrophic brain in the elderly patients facilitating the progression of the epidural hematoma because of lesser tamponading effect. He could find only one earlier report of epidural hematoma with Paget's disease of skull.^[2] There are reports of advanced Paget's disease with acute extradural hematoma after a trivial trauma.^{[4],[5]}

Extradural hematoma of the vertex has been relatively uncommon. There are quite a few single case reports and a few small series of cases.^[6-11] In pre-Computerized Tomography (CT) and pre-Magnetic Resonance Imaging (MRI) era the diagnosis of these hematomas were difficult and could be done only with venous phase of angiogram. The first vertex extradural hematoma diagnosed with MRI was reported by Ramesh and Sivakumar.^[10] Coronal CT also makes the diagnosis easier as in the present case, compared to the axial CT. Most of these hematomas have been due to injury to the superior sagittal

sinus. But in the present case, there has been no injury to the superior sagittal sinus.

In the present case, there was no skull fracture or injury to the superior sagittal sinus. The possible mechanism for the formation of extradural hematoma appears to be the bleeding from the inner table of the highly vascular Pagetic skull. The cause for subsequent deterioration after evacuation is evidently the acute venous hypertension due to pressure on the superior sagittal sinus, caused by continuous oozing from the Pagetic skull. Removal of the diseased skull flap controlled the collection of extradural hematoma. This is the first reported case of vertex extradural hematoma occurring in association with Paget's disease of skull.

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