

Sarcomatous Change in Polyostotic Paget's Disease-A Diagnostic Conundrum

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ABSTRACT

This is a report of a 56-year-old man presented to highlight the clinical features and diagnostic difficulties of Paget's disease in our environment. The patient had a four year history of intractable body pains and at different times had pathological fracture, loss of vision, conductive hearing loss, frequent intermittent anaemia requiring blood transfusion, paraparesis, one sided headache, cardiac failure and suspected Foster Kennedy syndrome. An earlier bilateral orchidectomy was done for suspected metastatic carcinoma of the prostate following markedly elevated acid phosphatase and ESR in the presence of enlarged hard prostate but a trucut biopsy of the gland later showed no evidence of malignancy. A multidisciplinary clinical assessment along with sequential ancillary test, roentgenography and bone marrow study helped to arrive at the eventual diagnosis (*Nig J Surg Res 2000; 2:88-91*)

KEY WORDS: Paget's disease, Sarcomatous change

Introduction

Paget's disease or osteitis deformans is a metabolic disease of bone characterized by marked increase in osteoblastic and osteoclastic activity with consequent increased bone turnover. It is thought to be rare in Asia, Africa and the Middle East.¹ The increased activity of bone results in unusual brittleness while the alternating osteolysis and osteosclerosis leaves a characteristic pattern on roentgenography.^{1, 2}

The disease may affect one bone (monostotic) or multiple bones (polyostotic). Deformities develop in the long bones of the lower limbs giving the patient an ape-like appearance.^{1,2}

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There may be pain, enlargement of the skull with platybasia and signs of cranial nerve compression leading to impaired vision, deafness and trigeminal neuralgia. Spinal cord or nerve root compression may result from vertebral thickening.¹⁻³

Complications arising from Paget's disease make diagnosis difficult even though the characteristic X-ray appearance may be pathognomonic.^{1,2} The various manifestations may mimic intracranial lesions, osteoarthritis, metastatic bone disease, primary cardiac conditions or multiple myeloma.^{1,2} This case is presented to illustrate the diagnostic difficulties encountered in Paget's disease in addition to its complications and progression.

Case report

A 56 years old man was referred to the department of surgery of Jos University Teaching Hospital on request of convenience in August 1997. He had persistent low back pain that radiated to the lower limbs. The pain was constant and unrelieved by analgesics. In addition, he had left shoulder pain secondary to pathological fracture, bilateral hip joint pains and generalized body aches of four years duration. He had a stoop and walked slowly with a walking stick. He had full power in all limbs and had no neurological deficits. There were no constitutional or urinary symptoms. Roentgenography revealed healing pathological fracture of the surgical neck of the left humerus and osteophytes of the lumbar vertebrae and acetabular margins. Osteosclerotic lesions were seen on L4, L5 and head and neck of the left humerus. There were alternating sclerosis and rarefaction of the bones of the skull, pelvis and proximal part of the left

humerus with distinct marrow/cortical demarcation.

Alkaline phosphatase level was high at 192 IU/L (Normal = 21-92 IU/L). Erythrocyte sedimentation rate (ESR) was 158mm/hr (Westergren Method) and there was a relative lymphocytosis of 43% and white blood cell (WBC) count of 6000/mm³. There was anaemia with packed cell volume of 24% for which he was transfused two units of blood. Mantoux test was 8mm. There was no Bence Jones protein in the urine.

A working diagnosis of polyostotic Paget's disease of bone with pathological fracture was made. While the patient was being investigated on presentation, the following differentials were entertained, viz: Lumbar spondylosis, tuberculosis of the spine, metastatic bone lesions and multiple myeloma.

Past medical history later revealed that he had been treated two years earlier by the referring hospital for suspected metastatic carcinoma of the prostate gland. At that time, he was said to have had his low back pain triggered off by trauma to the back in a road traffic accident four years previously. This progressed within four months until the patient was unable to stand. He had marked tenderness of the lumbosacral region. The power in the lower limbs were 2/5 and 1/5 on the right and left respectively. The prostate gland was said to have been enlarged and had a hard consistency. Pelvic radiograph was said to have shown osteolytic lesions of the pelvis and lumbosacral region. The ESR was 130mm/hr (Westergren method), acid Phosphatase 39 IU/L (Normal= less than 31 IU/L) and Bence Jones protein was negative. He had bilateral orchidectomy and was treated with stilboesterol, non-steroidal anti-inflammatory drugs (NSAID) and

physiotherapy. He regained function in the lower limbs and was able to ambulate but the pains persisted until he presented at Jos University Teaching Hospital.

A transperineal trucut biopsy of the prostate gland showed fibrocollagenous tissue only without evidence of malignancy. A combination of NSAID and physiotherapy was not helpful in reducing the pains.

Two months later the patient developed sharp, right-sided headache with diplopia and sudden loss of vision in the right eye. Ophthalmologic review entertained a diagnosis of demyelinating disease and lateral rectus paralysis while review by physicians suggested that the patients might be having Foster-Kennedy syndrome. He had two further episodes of severe anaemia at three monthly intervals requiring blood transfusion. He developed anaemic heart failure and recovered after treatment with frusemide and further blood transfusion. A bone marrow aspiration biopsy was done and the result showed hypercellularity with severely depressed erythropoiesis, megakaryopoiesis and myelopoiesis. The bone marrow was reported to have been infiltrated with clumps of malignant cells. No plasma cells were seen. A diagnosis of metastatic deposits to the bone was returned and bone biopsy advised.

After nine months of initial presentation, the patient developed loss of hearing in the left ear. Otorhinolaryngological review gave a diagnosis of conductive deafness in that ear. A wedge bone biopsy was taken from the right iliac crest where visual observation showed abnormal bone. The histopathology report was that of normal bone tissue without evidence of malignancy. The patient's symptoms continued exacerbating. He had five admissions for acute relapse of pains and

in order to have blood transfusion for anaemia. The patient later left the hospital without discharge. A report reaching us from his relations is that he has since died.

Discussion

The clinical and laboratory findings in osteitis deformans varies according to the stage of the disease.^{1,2} It would appear as if the bilateral orchidectomy offered to this patient abinitio at the peripheral hospital was too hasty. The initial treatment for presumptive carcinoma of the prostate, involving bilateral orchidectomy and oestrogen therapy was done without the benefit of histology, which is conclusive, despite suggestive but deceptive clinical signs and results of investigations. Empirical treatment is usually not accepted in the treatment of carcinoma of the prostate where the appearance of many lesions can mimic the disease on radiographs. The osteolytic lesions seen on X-ray may have called for caution since prostate deposits usually produce osteosclerotic lesions in bone. It would not be ruled out that trucut biopsy of the prostate may not give a representative specimen since only one or two cores of tissue are taken instead of the recommended sextant biopsy.^{4,5} The improvement achieved by the initial treatment at the peripheral hospital may have resulted from physiotherapy, NSAID, and temporary cessation of active disease. The lumbar spondylotic changes may have contributed to the symptoms experienced by the patients.

The pathological fracture of the humerus and later- x-ray changes in the skull, pelvis and humerus in addition to the alternate elevations of acid phosphatase and alkaline phosphatase favour the diagnosis of Paget's disease.^{1, 2,3} A high index of suspicion is

however required in early disease when changes are equivocal.³

The frequent anaemia and markedly elevated ESR are indices of sarcomatous change and make screening for multiple myeloma imperative. In this patient, Bence Jones protein was consistently negative, both at the referring hospital and on presentation. The loss of vision, conduction deafness and cardiac failure suffered by this patient are known complication of the disease.^{1,2}

The histopathological result does not exclude the diagnosis of sarcomatous change in Paget's disease as bone specimen are generally known to present histological diagnosis difficulties. However, a diagnosis made on the basis of clinical presentation, laboratory and radiological findings, progression of the disease, and bone marrow aspiration biopsy is no less authentic.

In conclusion, polyostotic Paget's disease presents diagnostic difficulties especially when complicated by early sarcomatous change and marrow infiltration.

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