Unilateral sclerosis of upper femur

A Kumar, M Shoaib

A 75-year-old man, a known patient with diagnosed prostate carcinoma for about 18 months, presented to us in the emergency department with sudden onset, excruciating pain around the right hip and thigh region. He was unable to walk or bear weight on the right side. There was no history of trauma or fever and he had never had similar pain in the past. He was on medical treatment for his prostate carcinoma. On examination he was in obvious discomfort from pain despite having an intramuscular analgesia. All the vital signs were stable. Local examination revealed an increase in skin temperature and tenderness in the upper thigh. Any attempts at palpation or movement aggravated the pain. There was no obvious swelling, abnormal mobility or crepitus. Routine blood investigations (blood counts, urea and electrolytes) were within normal limits. Bone profile was recorded as calcium 2.23 mmol/l (normal range 2.2–2.6), phosphorus 1.08 mmol/l (0.8–1.4), alkaline phosphatase 104 IU/l (40–120) and prostate-specific antigen 4.1 µg/l (0.0–4.0). An X-ray of the pelvis is shown in the figure. X-Rays of lumbar spine, chest and skull did not reveal any bony abnormality.

Department of
Trauma &
Orthopaedics, Dudley
Group of Hospitals,
Dudley, West Midlands
DY1 2HQ, UK
A Kumar
M Shoaib

Correspondence to
Dr A Kumar, 99 Lea House,
Russells Hall Hospital,
Dudley, West Midlands
DY1 2LU, UK

Accepted 10 March 1998

Figure X-ray of pelvis

Questions

- 1 What is the most probable diagnosis?
- 2 What investigation should be done to confirm the diagnosis?
- 3 What are the possible complications of the disease?

Answers

QUESTION 1

The most probable diagnosis is monostotic Paget's disease of the right femur. The other possible differential diagnosis are as listed in box 1.

QUESTION 2

A bone scan should be done to confirm the diagnosis and also to look for other areas of activity and pathological microfractures which are not detected on plain radiographs. If sarcomatous change is suspected, a biopsy should be done to confirm the diagnosis.

QUESTION 3

The possible complications of Paget's disease of long bones are pathological fractures, neoplastic degeneration and osteoarthritis of neighbouring joints.

Discussion

The clinical features and radiological findings (box 2) make monostotic Paget's disease of the right femur the most likely diagnosis. Acute pain in Paget's disease can be because of the acute stage of the disease itself, due to pathological fracture, sarcomatous degeneration, and in our patient, metastasis from prostate carcinoma. In the acute stage it typically presents as acute pain in the limb with local increase in skin temperature as a result of increased blood flow. The pathological fractures are present on the convex border of the bone, unlike Looser's zone in osteomalacia which are present on medial border. These fractures may not be detected on plain radiographs in which case bone scan is helpful. The sarcomatous degeneration presents as

Differential diagnosis of monostotic Paget's disease

- metastatic carcinoma of prostate
- lymphoma
- fibrous dysplasia
- myeloid metaplasia
- sclerosing osteitis of garre
- fluorosis

Box 1

Radiological features of Paget's disease of long bones

- areas of bone resorption
- areas of new bone formation
- irregular and coarse trabeculae
- increase in cortical thickness
- starts from one end and spreads to other
- V-shaped advancing end demarcates from healthy bone
- transverse incomplete or complete fractures on convex border

Box 2

Risk factors for neoplastic changes

- male > female
- > 60 years
- polyostotic disease
- involvement of long bones

Box 3

severe acute pain or an increase in chronic pain mostly associated with soft tissue swelling.¹⁻³ Sometimes pathological fracture is the first presenting complaint.

The risk factors for neoplastic degeneration in Pagetic bones are given in box 3. Radiologically, it appears as a lytic area in 68% of cases, mixed lesion in 26% and sclerotic in 6%,⁴ with soft tissue swelling. Unlike osteosarcoma of young adults there is hardly any periosteal reaction.

Carcinoma of prostate can spread to bones through blood and the bones commonly involved are vertebrae, femur, pelvis, ribs and sternum. Clinically, the presentation is severe bone pain and anaemia, in addition to a history of prostatism. Radiologically, these metastatic deposits are usually blastic or of mixed type with some evidence of bone destruction. The long bone looks expanded, with new bone formation. The occurrence of metastatic deposits in association with Paget's disease in the same bone is not very common but cases of adenocarcinoma,⁵ multiple myeloma,⁶ malignant giant cell tumour,⁷ and carcinoma vulva⁸ have been reported in the literature. There are very few reported cases of the occurrence of prostate carcinoma with Paget's disease in the same bone. Goldenberg (1961) mentioned a case of metastasis from prostate to Pagetic iliac bone.⁹ Due to the increased blood supply to the limbs in Paget's disease,¹⁰ there may be preferential haematogenous metastasis to the affected bone from a primary site.⁸

Paget's disease can be detected much earlier by a bone scan, which may show a monostotic Paget's disease to be a polyostotic disease. Bone scans help not only in diagnosis but also in assessment of the activity of the Pagetic lesion. The active lesion appears as a hot spot on the scan while inactive or sclerotic lesions may not be visualised. An increased uptake may also be seen in metastasis, fibrous dysplasia and osteitis but these can be differentiated on the basis of clinical features and plain radiographs. In sarcomatous degeneration, bone scan reveals increased uptake in the Pagetic bone but decreased uptake by the sarcomatous mass, most of which is necrotic due to rapid growth. In certain situations, such as pathological fracture or when bone scan is inconclusive, biopsy should be done to rule out sarcomatous degeneration.

The common complications of Paget's disease in long bones are pathological fractures, neoplastic degeneration, and osteoarthritis of neighbouring joints. The incidence of neoplastic degeneration as reported in the literature is usually slightly less than 1%.¹⁻³ Sarcomatous

changes can occur in the form of osteosarcoma, fibrosarcoma, lymphosarcoma and reticulum cell sarcoma, but osteosarcoma is the most common.¹⁻³ The prognosis in such cases is very bad with high mortality. Osteoarthritis of neighbouring joints occurs as a result of old age and abnormal mechanical stresses over the joint because of bent and deformed bones.

Progress

A bone scan was done and the findings were consistent with a diagnosis of Paget's disease in the right femur. It also revealed small areas of activity in the skull and pelvis. There was no

evidence of pathological fracture, sarcomatous degeneration, or metastasis from prostate. It was concluded that pain was due to an increase in activity of the Pagetic lesion in the femur. The patient was put on bed rest and analgesics to which he responded and he went home walking.

Final diagnosis

Polyostotic Paget's disease of bone.

Keywords: prostate carcinoma; femur; Paget's disease; sarcomatous degeneration; sclerosis

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Stroke in a young woman

I S Gambhir, D S Singh, D N Pattnaik

A 12-year-old girl from rural Varanasi was stung by a scorpion (*Palamnaes swannerdani*, India) on her right index finger. She developed intense local pain and swelling over the site, followed by unconsciousness and right-sided hemiparesis, without seizures, in one hour. There was no history of substance abuse or administration of sympathomimetic agents. On examination, 48 h after the sting, consciousness had improved but patient had right-sided hemiparesis, right supranuclear 7th nerve paresis with expressive aphasia. Examination of fundus and other systems was normal.

Total and differential leucocyte count, platelet count, haemoglobin, packed cell volume, erythrocyte sedimentation rate, bleeding time, coagulation profile, blood urea and sugar, urinalysis, chest X-ray, postero-anterior view, and electrocardiogram (ECG) on the day of admission were within normal limits. Her cranial computed tomography (CT) scan is shown in the figure. Carotid angiogram did not reveal any abnormality. Hemiparesis and aphasia improved during a follow-up period of two months.

Figure CT scan

Questions

- 1 What is the diagnosis?
- 2 What are the probable causes?

Department of
Medicine, Institute of
Medical Sciences,
Banaras Hindu
University, Varanasi -
221 005, India
I S Gambhir
D S Singh
D N Pattnaik

Correspondence to Dr IS
Gambhir, D 53/97 M7,
Parvati Puri, Guru Bag,
Varanasi, India

Accepted 10 March 1998