Case report

Ischaemic necrosis of the ilium complicating haemolytic anaemia due to an unstable haemoglobin

N W McGill, P Warburton, H Kronenberg, J E Hassall, and M A Rossleigh

From the Royal Prince Alfred Hospital, Camperdown, Australia

Summary A case of ischaemic necrosis of bone (INB) affecting the right hemipelvis in a 57 year old woman with an unstable haemoglobinopathy is presented. The rarity of INB in this site, the usefulness of nuclear scanning, and the relation between haemolytic anaemia and INB are discussed.

Key words: avascular necrosis, aseptic necrosis, osteonecrosis, haemoglobinopathy.

In adults ischaemic necrosis of bone most commonly affects the epiphyseal (especially the femoral and humeral heads) and metadiaphyseal marrow cavities of long tubular bones. Occasionally, the small round bones of the wrist and ankle are affected. Involvement of large flat bones such as the ilium is rare and to our knowledge has only previously been described complicating sickling diseases, Gaucher’s disease, and in two patients who received large doses of corticosteroids in the course of renal transplantation.

We report a case of ischaemic necrosis affecting most of the right hemipelvis in a 57 year old woman with haemolytic anaemia due to an unstable haemoglobin. Despite a lifelong history of haemolysis there was no evidence of previous episodes of ischaemic necrosis, nor did she have a sickling disorder.

Case report

A 57 year old woman presented in January 1987 with severe pain in the region of the right hip. She was diagnosed as having an unstable haemoglobinopathy at the age of 19 years when noted to be anaemic during her first pregnancy. Over the next 30 years she received only occasional blood transfusions but for the eight years before this presentation received transfusions of four or five units of packed cells approximately every six weeks. Her unstable haemoglobin was characterised by the presence of small numbers of Heinz bodies and numerous other inclusion bodies which have been shown to contain haemoglobin. Attempts in the early 1970s to analyse her haemoglobin using various forms of haemoglobin electrophoresis and peptide analysis (finger printing) were unsuccessful owing to the instability of her haemoglobin. Further attempts to characterise the haemoglobin, including amino acid sequencing, are currently being undertaken.

As a consequence of her chronic haemolysis she had a cholecystectomy for pigment stones in 1969 and a splenectomy for abdominal discomfort due to massive splenomegaly in 1973. She had noted increasing skin pigmentation for the five years before presentation.

Since 1975 she had suffered from a non-deforming polyarthritis which affected her hands, wrists, ankles, and shoulders. She had mild intermittent joint swelling and prominent early morning stiffness. Rheumatoid factor was negative and x rays of her joints were normal. Although she was initially thought to have seronegative rheumatoid arthritis, the subsequent course of her arthritis was more in keeping with the arthropathy of iron overload which she was recorded to have. In 1985 she developed transient pain in the region of the right hip. A bone
scan was normal (Fig. 1), and no further investigation was considered necessary. In 1986 she suffered an osteoporotic crush fracture of T11.

She also had a number of intercurrent illnesses including peptic ulceration, hypertension, ischaemic heart disease, and aortic incompetence.

She was a non-smoker, drank minimal alcohol, and took isorbide nitrate, nifedipine, pindolol, ranitidine, folic acid, indomethacin, paracetamol-dextropropoxyphene, and temazepam. She had never taken corticosteroids.

In January 1987 she developed over a few hours severe pain in the region of her right hip, which prevented her from walking and required large amounts of narcotic analgesia to achieve adequate pain relief even while lying in bed.

On examination she was distressed by pain and had a low grade fever to 37.8°C, which persisted for five days. Her right iliac crest was exquisitely tender, but movements of the right hip were only moderately restricted by pain. Her bones and joints were otherwise normal. Neurological examination was difficult because of her pain and subsequent sedation but was felt to be normal apart from a small area of longstanding hypoaesthesia on the lateral aspect of her right thigh. She had slate grey pigmentation, a liver edge palpable 3 cm below the costal margin, and signs consistent with moderate aortic incompetence.

Investigations at that time were haemoglobin 90 g/l, white cells $1.72 \times 10^9/l$, platelets $181 \times 10^7/l$. Liver function tests had been and remained chronically elevated.

Fig. 1 Technetium-99m methylene diphosphonate bone scan: normal; November 1985.

Fig. 2a

Fig. 2b

Fig. 2 (a) Technetium-99m methylene diphosphonate bone scan: reduced tracer uptake in the right ilium, most marked in the iliac crest and extending into the acetabulum; January 1987. (b) Technetium-99m sulphur colloid bone marrow scan: absent bone marrow uptake throughout most of the right hemipelvis; January 1987.
Ischaemic necrosis of the ilium

Fig. 3a  Fig. 3b

Fig. 3  (a) Technetium-99m methylene diphosphonate bone scan: persistence of reduced tracer uptake in the right ilium, only marginally less marked than in January 1987 (Fig. 2); March 1987. (b) Technetium-99m sulphur colloid bone marrow scan: persistent absence of bone marrow uptake throughout most of the right hemipelvis, unchanged from January 1987 (Fig. 2); March 1987.

abnormal (alkaline phosphatase 179 U/l, aspartate aminotransferase 76 U/l, alanine aminotransferase 83 U/l, γ-glutamyltransferase 46 U/l), but the biochemical screen was otherwise normal. An x ray of the pelvis showed some generalised loss of bone density but was otherwise normal. A ⁹⁹ᵐTc methylene diphosphonate bone scan showed reduced tracer uptake in the right ilium, most marked in the iliac crest and extending into the acetabulum (Fig. 2a). In view of the bone scan appearance a ⁹⁹ᵐTc sulphur colloid bone marrow scan was performed and showed absent bone marrow uptake throughout most of the right hemipelvis—the abnormality being more extensive than that present on the bone scan (Fig. 2b). The combined studies strongly supported the diagnosis of bone infarction. A computed tomographic scan of the abdomen and pelvis showed replacement of the normal fatty marrow in the femoral head consistent with increased haemopoiesis, and increased liver density consistent with iron overload, but no mass or bony erosion.

The patient was treated with rest and analgesia and improved rapidly, being able to walk again 11 days after the onset of pain. Unfortunately, troublesome low grade right pelvic pain persists eight months after onset.

Repeat bone and bone marrow scans performed after two months showed little change (Figs 3a and b) and pelvic x ray after seven months was unchanged. A trephine biopsy of the right ilium confirmed the presence of osteonecrosis (Fig. 4).

Fig. 4  Trephine biopsy of the right ilium. Absent osteocyte nuclei indicate necrotic bone trabeculi. The marrow spaces revealed numerous siderophages, sparse haemopoietic cells, and a few fibroblastic cells. (Haematoxylin and eosin.)


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Discussion

We have presented a case of ischaemic necrosis of bone (INB) occurring in an unusual site in a patient with an unstable haemoglobinopathy.

INB has been reported in association with a number of conditions (traumatic disruption of the blood supply, sickle cell disease and other sickling disorders, corticosteroid treatment, Cushing’s disease, irradiation, alcoholism, Gaucher’s disease, caisson disease, haemophilia, rheumatoid arthritis, systemic lupus erythematosus, and pancreatitis) but not to our knowledge with non-sickling haemoglobinopathies.4

Ischaemic necrosis of the ilium is rare but has been reported in sickle cell disease,1 Gaucher’s disease,2 and after large doses of corticosteroids for renal transplantation.3 In sickle cell disease ischaemia is thought to result from sludging of sickled red cells in small vessels and vascular sinusoids,4 and in Gaucher’s disease ischaemia may be due to compression of vessels as a result of increased intraosseous pressure resulting from infiltrating Gaucher cells.5 The blood supply of the ilium comes via the iliac branches of the obturator and iliolumbar arteries (branches of the internal iliac), which anastomose with each other and with branches of the superior gluteal artery (branch of the external iliac) to give off numerous small vessels which supply the bones of the pelvis.5 6 As occlusion of a major vessel could not account for ischaemic necrosis of the bones of the pelvis without also producing visceral ischaemia it is not plausible that our patient’s illness resulted from such an occlusion.

Our patient does not have a sickling disorder nor any of the other diseases associated with INB mentioned above. Although our patient’s bone marrow was hyperplastic, INB is not associated with other non-sickling haemolytic anaemias with similar degrees of marrow hyperplasia, and thus the relation between our patient’s unstable haemoglobinopathy and the occurrence of INB remains unclear.

This report describes INB occurring in an unusual site and demonstrates the value of bone and bone marrow scanning in patients with undiagnosed acute severe musculoskeletal pain and normal x rays.

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References

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