CASE REPORT

Pathological fracture of femoral neck in a middle-aged woman: a rare presentation of primary hydatid cyst disease in humans

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SUMMARY

Hydatid disease in humans is caused by *Echinococcus granulosus*. It most commonly involves the liver and, to a lesser extent, the lungs and spleen; however, it is known to involve other areas, too. Involvement of bone by hydatid cyst is rare. Here, we describe the case of a 37-year-old woman who presented with pain in the left groin and swelling in the left thigh. The radiological imaging showed a fracture of the femoral neck and cysts in the shaft of the femur. Diagnosis of hydatid cyst was confirmed on the basis of histopathology of biopsy specimens. The patient recovered after surgical excision of the cyst. This case illustrates the various sites and presentations of hydatid cyst disease, and the need to investigate for it if cystic bony lesions are encountered especially in endemic regions, as a delay in diagnosis can lead to long-term morbidity and even death.

BACKGROUND

Hydatid disease in humans is caused by four species of the *Echinococcus* parasite, the most common being *Echinococcus granulosus*.^{1–5} This disease occurs through the ingestion of parasite eggs from the ingestion of contaminated water or food.² Hydatid cysts have been known to develop anywhere in the body.^{2–3} They most commonly occur in the liver followed by lungs and the spleen.⁶ Hydatid cyst in the bone is very rare.^{4–9} It is endemic to parts of Asia, South America, Africa and Australia. The incidence of bone involvement is 0.5%–2% of all hydatid cyst infections.^{2–4–6–9–11}

Hydatid cyst disease in bone has a variable presentation and depends on the involved area. It is often asymptomatic but can occasionally present as pathological fractures and pain.^{2–4–6–12–13} Diagnosis of hydatid disease is based on the patient's clinical presentation, radiology and histopathology.^{1–4} Immunological tests and older investigations like the Casoni intradermal test are obsolete and no longer used.^{4–14–15} Radiology may be very non-specific; however, histopathological examination of the bone specimen is confirmatory.^{1–4–6–7–16–18}

Surgical removal of the cystic lesion is the main treatment.^{4–6} When bone is involved, the treatment is complete resection of the involved area with a wide healthy margin. The combination of antihelminthic therapy, wide resection and bone replacement with polymethylmethacrylate gives good

results.^{1–3} Prognosis of bone hydatidosis is variable and patients treated promptly usually recover; however, rarely, sepsis can occur and it may lead to death.^{1–2–6}

Here, we present a case of primary hydatid cyst disease of the femur without any primary hydatid cyst in the liver, which is an uncommon site for occurrence of this disease. The patient presented with pain in the left hip joint secondary to femoral neck fracture, which is also a rare presentation of hydatid cyst disease. Histopathology confirmed the diagnosis, and the patient was successfully treated with surgical resection and antihelminthic therapy.

CASE PRESENTATION

A 37-year-old woman presented with complaints of acute-onset dull and non-radiating pain in the left groin along with swelling of the left thigh. The patient had been having pain in her left hip and thigh for the last couple of months but the pain worsened significantly over the last 24 hours. The patient denied any trauma, fever or recent infection. Review of systems was negative except for the pain. There was no significant medical history other than the pain. The patient had previously visited a local general practitioner for the joint pain; however, no work-up was done at that time as the pain was mild. The patient belonged to a family of farmers and occasionally worked on the farms.

On examination, there was diffuse swelling in the left thigh with painful mobility in the left hip joint.

INVESTIGATIONS

Plain radiograph of the left hip joint and femur revealed a fracture of the neck of left femur and intramedullary cystic lesions of various sizes in the shaft of the femur (figure 1). There was cortical thinning of bone in the lower half of the shaft of the femur (figure 2). Tuberculosis and hydatid cyst were the most likely differential diagnoses.

Laboratory studies to identify the aetiology showed a normal complete blood count and an unremarkable erythrocyte sedimentation rate. Blood cultures were ordered but they did not reveal any growth after incubation. A purified protein derivative skin test did not result in any significant induration and a gamma interferon assay was also normal.

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Figure 1 Plain radiograph of the left hip joint showing fracture of the neck of the femur.

Surgical excision of the cyst was done, and the specimen was sent for histopathological examination. Microscopic sections revealed multiple fragments of lamellated bone. Benign lamellated acellular cysts were identified (figures 3 and 4). There was no evidence of acute or chronic inflammation, granuloma or malignancy in the sections examined. Thus, histopathological examination confirmed the diagnosis of hydatid cyst in the femur.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis before biopsy was done included

- ▶ Hydatid cyst disease
- ▶ Tuberculous osteomyelitis
- ▶ Chronic osteomyelitis
- ▶ Primary or secondary neoplasm
- ▶ Infiltrative bone disease
- ▶ Degenerative joint disease

TREATMENT

The patient was treated with surgical removal of all cysts, irrigation with chlorhexidine as a scolicidal solution, bone grafting into the bone defects and immobilisation of the fracture. This was followed by 3 months of oral albendazole.

OUTCOME AND FOLLOW-UP

The patient was followed for 6 months and remained asymptomatic.

DISCUSSION

Hydatid disease is caused by the larval stage of the tapeworm *E. granulosus*.^{1 2 19} It most commonly involves the liver and lungs but has been reported to develop in a variety of different

Figure 2 Plain radiograph showing cortical thinning in the shaft of femur and intramedullary cystic lesions in the lower half of the shaft of the femur.

locations in the body.^{1 2 6} Hydatid cyst in bone is very rare.^{6 7} When the bone is involved, the most common site is the spine, followed by the pelvis, femur, tibia, ribs, skull, scapula, humerus and fibula.⁴

The life cycle of *E. granulosus* involves two hosts.⁶ The definitive hosts are usually dogs and certain wild carnivores.⁶ The intermediate hosts are usually sheep, goats and other herbivores.⁶ Humans act as an intermediate host.¹ The ova ingested

Figure 3 Photomicrograph (H&E stain, (x40) of bone fragment showing hydatid cysts (arrows)).

Figure 4 Photomicrograph (H&E stain x100) showing hydatid cyst. The cyst comprises acellular lamellated membrane (arrow).

by humans penetrate the intestinal wall, reach the portal system and then the liver.^{1,20} A few ova may pass through the liver and reach the lung, and then into the systemic circulation, which explains the occurrence of hydatid disease in other organs.¹ Dissemination through lymphatic channels may be responsible for hydatid cysts at uncommon sites.^{1,20}

Hydatid cyst disease in the bone has a very variable presentation and is often asymptomatic.^{2,4} The bony lesions may be in the form of solitary or multiple cysts.^{4,5} They may be detected after a pathological fracture or an infection.^{2,4,6,12,13}

The diagnosis of hydatid cyst disease in the bone is quite challenging.^{2,10} Diagnosis of hydatid disease is based on the patient's history, clinical findings, radiological examination and histopathology.^{1,4} Older tests like the Casoni intradermal test are no longer used because of its high rate of false-positive results.^{4,14} Serology can be used for both the initial diagnosis and for the follow-up treatment of *E. granulosus*.^{15–17} Numerous antibody and antigen tests are available. However, their use is usually limited by their sensitivity. There are many pitfalls associated with serological testing. A negative serological test especially in a patient with high suspicion does not rule out hydatid cyst disease. Their results also depend on a lot of factors including the location and viability of the cysts. Besides this, the tests do not correlate with disease burden either. However, they do offer a non-invasive way to diagnose hydatid cysts and sequential testing with more than one serological test has been shown to improve sensitivity.¹⁸ Studies have shown ELISA to be the most sensitive, ranging from 80% to 92% in patients with liver disease. IgG is more sensitive than IgM. This is followed by haemagglutination, immunoblot (using antigen 5 and/or a B-rich fraction) and enzyme-linked immunotransfer blot, all of which were 80% sensitive. Latex agglutination had a sensitivity of 65%–75%. These tests have a lot of variability in their sensitivity. This is due to different laboratories using different reference ranges. This is compounded by the fact that serological tests also vary in their sensitivity based on the organ system involved. In general, antibody tests are more sensitive than antigen tests.^{4,19–23} Radiological imaging studies, although non-invasive, are not very specific for hydatid cyst disease.^{1,2} The most common radiological manifestation of bone hydatid disease is a lucent expansile lesion with cortical thinning.^{3,4,6,24} The findings of X-ray or CT scan are usually similar to those of tuberculosis, metastases and giant cell tumour.^{4,24–28} It has to be confirmed by histopathological examination of the bone specimen.^{1,4,6} On histopathological

Learning points

- Hydatid cyst in bone is a rare phenomenon and can pose a diagnostic challenge.
- It should be considered as a differential in patients with unexplained chronic bone pain especially in endemic areas.
- Diagnosis can be achieved by imaging followed by confirmation with a bone biopsy.
- A delay in diagnosis can result in extensive bone involvement with resultant surgical removal causing long-term morbidity.
- Treatment involves surgical resection with bone grafts if required, complement with oral albendazole therapy.

examination, the cyst external wall is chitinous and internally the germinal layer is surrounded by a fibrous capsule and granulation tissue.²⁹

There are no specific guidelines on how to treat hydatid cyst disease of the bone, but treatment usually involves surgical resection of the bone along with oral antihelminthic therapy. Surgical resection is accompanied by irrigation of the cystic pouch with a scolical agent like silver nitrate, hypertonic saline or iodine solution. The defects produced by resection can be filled with a bone graft or synthetic material like polymethylmethacrylate. This should be accompanied by antihelminthic therapy like albendazole for up to 3 months postoperatively to prevent recurrence of the disease.^{30–34}

Early diagnosis of bone hydatidosis helps in eradication of cyst. Misdiagnosis or delayed diagnosis may lead to amputation, recurrence and sepsis.^{2,4,7}

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