

Hydatid Disease of The Bone: A Rare Presentation

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ABSTRACT

Hydatid disease of the bone is a very rare condition. It represents about 0.5–4% of all human hydatid disease. Spine is the most affected part of the skeleton (50% cases). Extra-spinal bone hydatidosis is much rare. A 20 year old woman presented with severe pain and swelling in left thigh. On examination, there was a boggy swelling in left thigh suggestive of abscess. MRI of left hip joint and thigh revealed features of osteomyelitis with pathological fracture in upper shaft along with multiloculated collections in upper thigh muscle. Biopsy revealed evidence of Hydatid cyst. Hydatid disease of the bone is a rare manifestation of Echinococcosis. It is hence important to consider the possibility of Hydatid disease of the bone as a differential diagnosis of lucent lesions of bone especially in areas where it is prevalent for proper treatment of the patient. Radiological, laboratory, and clinical findings combined with strong element of suspicion are the key for diagnosis.

Keywords: Bone, Echinococcus, Hydatid cyst, Recurrence

Introduction

Hydatid disease is caused by the larval stage of the Echinococcus tapeworm. It can occur almost anywhere in the body with most common sites being liver and lung.¹ Hydatid disease of the bone is a very rare condition representing about 0.5–4% of all cases.^{2,3} Spine is the most commonly affected part of the skeleton (50% cases). Extra-spinal bone hydatidosis is much rare.⁴ Osseous hydatidosis is difficult to treat and carries high morbidity due to frequent recurrences, especially in certain locations such as ilium and hip, where radical surgery is difficult. We report a case of hydatid disease of femur presenting with a hairline fracture complicated by chronic osteomyelitis and sinus formation with multiple recurrences. In view of rarity of the disease at this site with complications and frequent recurrences evokes an interest in this case.

Case Report

A twenty-year old woman presented with severe pain and swelling in left thigh of eight months duration. On examination, there was a boggy, indurated swelling in the left thigh suggestive of an abscess. X-Ray and MRI of left hip joint and thigh revealed features of osteomyelitis with extensive cortical and marrow destruction, suggestive of cystic content/abscess formation. It also showed a pathological fracture in the sub trochanteric area with lateral displacement of distal femur shaft with contiguous multiloculated collections in upper thigh muscle (Figure 1). Hematological parameters were normal except for mild anemia. The patient underwent surgical debridement at a

private hospital, started on antibiotics and was immobilized. This was followed by a second surgery four months later wherein curettage was done and sent for histopathological examination and an interlocking intramedullary nail was placed. Biopsy revealed evidence of Hydatid cyst comprising of cyst wall showing lamellations with multiple areas of calcification. Few brood capsules with hooklets were seen. The bony tissue pieces showed palisading layers of inflammatory cells with numerous multinucleated giant cells. The outer parts of the pericyst showed newly laid down bone (Figure 2). No lesions were detected in any other organ by imaging including chest X-Ray and abdominal ultrasound. There was no history of anaphylactic reaction after the fracture. The post-operative period was uneventful and she was discharged on appropriate anti helminthic therapy (Albendazole) and advised bed rest. After three years, she came back with complains of continuous pus like discharge from the suture area, increased swelling in thigh, shortening of leg and partial weight bearing. The clinical possibility considered was superadded infection or tumor. The patient underwent diagnostic incisional biopsy which revealed evidence of hydatid disease recurrence. She was started Albendazole following which she remained asymptomatic for three months after which she again developed swelling and discharge. CT scan of the hip joint showed multiple osseous and soft tissue cysts with diffuse cortical thickening and irregularity involving most of the femur and a thick fistulous tract in lateral thigh suggestive of diffuse osseous and soft tissue hydatidosis with chronic osteomyelitis of femur and thereby indicating

the spread of infection to both hip and knee joints making a radical surgery difficult. She was again started Albendazole and kept on immobilization for two months. Following this pain and swelling reduced, however there was no improvement in discharge. In recent follow-up, CT scan showed similar picture as before with increased periosteal reaction. It revealed persistence of the intramuscular cysts with increase in size.



Fig. 1: X-Ray of left hip joint and femur showing pathological fracture in upper shaft with lateral displacement of distal femur.

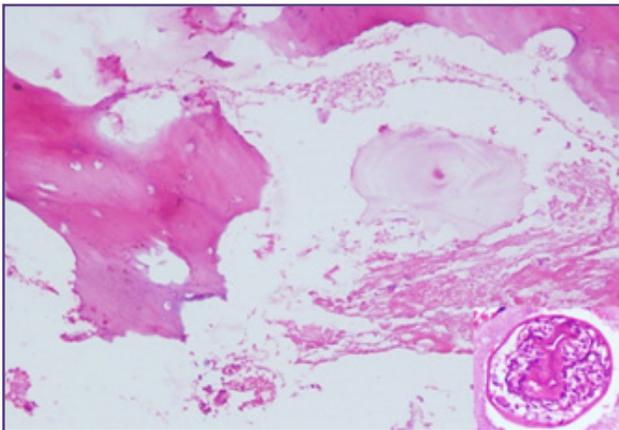


Fig. 2: Section shows cyst wall having lamellations within the bony trabeculae. The outer parts of the pericyst shows newly laid down bone. Inset shows a brood capsule with hooklets. (H&Ex400).

Discussion

Hydatid disease is a parasitic infestation produced in the intermediate host like man, sheep or cattle by the tapeworm *Echinococcus*. *Echinococcus granulosus* is the most common cause of hydatid disease in humans. The disease spreads when food or water that contains the eggs of the parasite is ingested or by close contact with an infected animal.^{5,6} The commonest sites affected

are liver and lungs as they act as first and second filters respectively.⁷ Hydatid disease of the bone is a very rare condition. Spinal hydatidosis can be primary or secondary from extraspinal hydatidosis through protovertebral shunts or direct extension.⁸ The frequency of osseous involvement in hydatid disease is 0.5%–4%.^{2,3} Spine is the most commonly affected part of the skeleton (50% cases). Extra-spinal bone hydatidosis is rare and may affect pelvis, femur, tibia, humerus, skull, and ribs.⁴

Osseous foci may be manifested as pain and deformity, particularly in 30-60 years old age group. Hydatid disease of bone is rarely seen in childhood.⁹ It remains asymptomatic over a long period (as long as 40 years) due to high resistance of bone and is usually detected when lesions have become extensive like after a pathological fracture, neural deficit, secondary infection, fistula formation or following the onset of compressive myelopathy in cases of vertebral lesions. Therefore, management is difficult and recurrence is common.¹⁰

Skeletal cystic echinococcosis lesion may be single or multiple. The initial location of the lesion in long bones is metaphyseal or epiphyseal, later extending to the diaphysis.¹⁰ In bone involvement, pericyst formation does not occur, thereby allowing aggressive proliferation in an irregular branching fashion along the line of least resistance, especially the bone canals.⁴ The parasite replaces the osseous tissue between trabeculae due to the slow growth of multiple vesicles. With time, the parasite reaches and destroys the cortex, with subsequent spread of the disease to surrounding tissues, at times, leading to calcification.^{4,11}

The X-ray and CT scan appearances are non-specific with most common being a lucent expansile lesion with thinning of the cortex.¹² The differential diagnosis of skeletal cystic echinococcosis includes other infectious lesions like tuberculosis, chronic osteomyelitis, fibrous dysplasia and certain tumors like simple bone cyst, aneurysmal bone cyst, plasmacytoma, osteosarcoma, chondrosarcoma, chondromyxoid fibroma, lymphoma, giant cell tumors, brown tumor, metastases, etc.^{4,13,14}

Pre-operatively, definitive diagnosis of bone hydatidosis is often difficult. Radiologic and serologic findings can generally help establish the diagnosis, but unusual location with atypical imaging findings may complicate the differential diagnosis. The diagnosis is difficult since daughter cysts, calcification, and germinal membrane detachment, typical manifestations of cystic echinococcosis in parenchymal organs, are not usually observed in skeletal hydatid cysts. An accurate diagnosis of hydatidosis may be aided in some cases by eosinophilia (25 to 35% of cases). Casoni intradermal test and Weinberg complement

fixation test were former diagnostic tools, which have been disused due to false positive results and lesser sensitivity in extrahepatic hydatidosis.¹⁵ Histopathological examination is considered gold standard for a definitive diagnosis.¹⁴

In bone, the lamellated layer of hydatid cyst is not well developed and lesion migrates along the canaliculi of bone like as a metastatic growth. Secondary infection plays an important role in killing the hydatid parasite. Pathological fracture of long bone due to primary hydatid cyst may be complicated by localized secondary echinococcosis, non-union, sinus formation and sometimes anaphylactic reaction.

Treatment of osseous hydatidosis is closer to oncologic therapy than to the usual surgical treatment of visceral hydatid cysts. Because of the poor results with medical treatment, surgery is the treatment of choice for hydatid bone lesions. Many authors have advocated wide resection of the involved bone along with surrounding soft tissues as the only definitive treatment of the condition with or without chemotherapy using albendazole or mebendazole.^{7,10} The prognosis of osseous hydatidosis remains poor, especially with spinal and pelvic localizations, which are the most frequent ones. The prognosis and treatment of osseous hydatidosis belong in the same category as a locally malignant lesion.

In the present case, patient might have been harbouring asymptomatic infestation for quite a long time and it was only pain that brought her to us for treatment. Further the management was complicated by an extraosseous involvement, cortical erosion, chronic osteomyelitis and pathological fracture.

Conclusion

Hydatid disease of the bone is a rare manifestation of Echinococcosis. There is insidious progression of the parasite into bone, leading to an immediate diffuse, extensive, invasion process with fracture. The treatment of osseous hydatid disease is entirely surgical with or without chemotherapy, however recurrence is likely. It is hence important to consider the possibility of Hydatid disease of the bone as a differential diagnosis of lucent lesions of the bone especially in the areas where it is prevalent; so as to render proper treatment to the patient. Radiological, laboratory, and clinical findings combined with strong element of suspicion are the key for diagnosis.

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