Case report

A healed echinococcus pathological femoral shaft non-union—A case report and a literature review

Josh E. Schroeder a,b, Yoram A. Weil a, Hila Elinav b, Rami Mosheiff a, Amal Khoury a,*

a The Orthopedic Surgery Department, Hadassah Hebrew University Medical Center, Jerusalem, Israel
b The Microbiology Department, Hadassah Hebrew University Medical Center, Jerusalem, Israel

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1. Introduction

Hydatid disease is prevalent throughout much of the world, mainly in the developing world and unprivileged areas. There are twelve subtypes of Echinococcus known today. However, in humans there are two main types of infection—Echinococcus multilocularis causing alveolar and Echinococcus granulosus which causes the cystic echinococcosis. The incidence of bone disease, caused by echinococcus is rare as only 0.5–4.5% of cases of echinococcosis have bony involvement. Primary hydatid disease of bone, is caused by E. granulosus, occurs when a blood-borne scolax settles in bone. In bone involvement, pericyst formation of the echinococcus does not occur, thereby allowing aggressive proliferation in an irregular branching fashion along the line of least resistance, especially the bone canals. When this proliferation causes damage severe enough to create a compromise in the solidity of the bone, a pathological fracture may occur. However, very few reports deal with the treatment of pathological long bone fractures, especially as the sole presentation of the echinococcus usually with a grim outcome. We present a case of a pathological femur fracture non-union caused as an isolated bony infection of echinococcus, treated successfully with serial debridements, antibiotics and fixation with a locked plate construct.

2. Case report

A previously healthy 61-year-old female presented at our clinic due to pain and inability to bear weight. She was a housewife, lives in a rural community, with a livestock of sheep and dogs, in an endemic area for echinococcus. Two years prior to her visit, she fell down and broke her femur. She underwent surgery in order to stabilise her fractured distal femur with a dynamic compression plate. A year later she was reevaluated and was diagnosed as suffering from non-union of the femoral fracture. Revision surgery was performed including re-fixation of the femur with a dynamic condylar screw system and autologous iliac crest bone grafting of the non-union sight. The patient presented in our clinic 1 month later with erythema, pain and drainage from the operative wound.

The patient was admitted and underwent debridement of the surgical wound and fracture site and removal several loose screws. The fixation plate and the condylar screw were retained in order to provide stability of the fracture, so the infection could be healed. The patient's treatment with vancomycin (one gram twice daily) and rifampicin (600 mg three times a day) was renewed. The patients' lower extremity was placed in a long hinged knee brace and weight-bearing was allowed. However, no cysts were seen in pathology. The anti-staphylococcal antibiotics were substituted for vancomycin (1 g twice daily) and rifampicin (600 mg three times a day). Three weeks postoperatively, the patient's pathology results showed echinococcal cysts. The patient's treatment was expanded albendazole (200 mg three times a day) and praziquantel (100 mg three times a day) were added as well. The patient's serology (indirect haemaglutinin) was positive for echinococcal infection. Total body imaging (CT scan and bone scan) did not demonstrate additional sites of cyst formation. The patient was discharged for home care continuing antibiotic and anti-helmintic care until a secondary procedure of removal of the cement spacer and eventual re-fixation will be performed. Three months postoperatively, a new secreting sinus appeared in the right thigh. The patient was admitted and underwent further debridement, removal of the gentamycin beads and existing hardware and placement of a long 4.5 mm titanium distal femoral locked compression plate (LCP) (Synthes, Battlach, Switzerland). The patient's lower extremity was placed in a long hinged knee brace and weight-bearing was allowed. However, no cysts were seen in pathology. The anti-staphylococcal treatment with vancomycin (one gram twice daily) and rifampicin (600 mg three times a day) was renewed. The patients' lower extremity was placed in a long hinged knee brace and weight-bearing was allowed.
bearing on the affected extremity was restricted. The patient was discharged for home care continuing the antibiotic and anti-helmintic care for 3 further months. Two months after the last procedure, the patient’s pain subsided, and there was radiographic evidence of fracture healing. Progressive weight bearing on the right lower extremity was initiated. One and a half year postoperatively, the patient had a leg to leg discrepancy of 5 cm and knee flexion is limited to 80 degrees but no pain on weight bearing and does not suffer from any functional limitation. X-rays obtained at that time demonstrated complete union of the fracture (Fig. 4).

3. Discussion

We report here a case of an isolated skeletal echinococcal disease, presenting as a non-union of a femur fracture. It was treated by debridement, antibiotics and rigid fracture fixation resulting in a solid healing of the femur.

Echinococcus is the most commonly granulosis disease in man. It mainly causes cystic echinococcosis (CE). However, osseous CE is the least common presentation of CE—and even in endemic regions of echinococcus it is rarely recognised as a cause of pathological fracture.

Primary echinococcal skeletal infections occur mainly in highly vascularised bones. The vertebrae, long bone epiphyses, ilium, skull, and ribs are most frequently affected. The parasite replaces the osseous tissue between the trabeculae with slow growing multiple vesicles. Afterwards, the parasite reaches and destroys the cortex, and spreads to surrounding tissues. The hydatid cysts may lie dormant in the bone for extended periods of time.

Most patients, as well as the one presented here, usually present with pain, swelling or pathological fracture. If the infection is in the spinal, it might manifest as severe back pain, or as weakness and sphincter disturbances due to cord compression. The differential diagnosis is wide and includes tuberculosis spondylitis, chronic osteomyelitis, aneurismal bone cysts, giant-cell tumours, solitary cysts, neurofibromatosis, fibrocystic disease, chondrosarcoma and tuberculosis. A detailed patient history must be taken and any risk factors must be noted (i.e. living in endemic areas, contact with live stock or dogs should be documented). The diagnosis of echinococcal infection in many cases is not clear and the medical teams caring for the patient need to embrace a high index of suspicion when caring for a patient with a infected non-union of a fracture.

When the clinical suspicion of echinococcal infection is high, serological assays might assist in making the diagnosis, however serology in osseous CE is less specific, indirect
haemagglutination (IHA) test (sensitivity 81.8%, specify 47%),\(^4\) counter immune-electrophoresis (CIE) (sensitivity 75%, false positive 25%),\(^3\) enzyme-linked immune-sorbent assay (ELISA) (sensitivity 90%)\(^1\)\(^1\) indirect immunofluorescence test (IFAT) (sensitivity 90%)\(^2\)\(^2\) and gold-labelled antibodies (sensitivity 90%).\(^2\)\(^1\) The two most common tests used are ELISA and IFAT for the diagnosis of osseous CE.

Adequate imaging must be utilised in order to confirm the diagnosis. Unfortunately, distinct radiographic features of this disease are not common. X-rays with osseous changes are found only in 27% of cases and “moth eaten areas” with surrounding sclerosis are typical.\(^1\)\(^0\) On CT, skeletal cystic hydatidosis appears as one or several closely related, well-defined, osteolytic lesions. There may be bone expansion, cortical thinning, cortical destruction, sclerosis, honeycomb appearance, and extension into adjacent soft tissues.\(^2\) The most helpful technique for diagnosing hydatid cyst disease is magnetic resonance imaging (MRI).\(^2\)\(^3\)

The treatment of osseous hydatid disease is primarily surgical; aiming to remove the cysts, surrounding bones and contaminated soft tissues, complete removal of the cyst is needed to avoid further dissemination of CE topical treatment with anti-helmintic care may assist with the eradication of the infection. With replacement of bone defects with bone grafts or prosthesis, the treatment goal is to achieve a stable and viable construct allowing proper healing of the bone and soft tissues. A secondary goal is avoidance of secondary infections, and to prevent the recurrence of the cysts, this is achieved with a topical cysticidal agent, usually albendazole.

Fig. 3. Histology. (A) 100× magnification of the tissue taken from the pathological fracture. Echinococcal cysts can be seen next to newly formed bone. (B) 400× magnification of the tissue, the echinococcus lamellar cyst can be seen and with the reactive inflammatory tissue and pathological new bone formation.

Fig. 4. The femur fixed with a LCP 4.5 supracondylar plate. A: AP view; B: lateral view of the femur. The fracture has healed.
Unfortunately, these achievements are rarely met in this relentless disease. Surgery is often only palliative. Prolonged treatment with combination of albendazole and praziquantel may improve the cure rate, in comparison to treatment with albendazole only (with less than 50% success rate), but causes more complications. Recently, treatment with nitazoxanide for bony lesions has been reported. However the success of any mode of cysticidal treatment is limited and more effective and tolerable chemotherapy is required.

Our case was complicated by a co-infection with MRSA. There is sparse information in the literature about the effects of a secondary infection. To the best of our knowledge, only one case of co-infection of echinococcus and MRSA was reported. In this case, a hip infection required an eventual total hip replacement. Unfortunately that patient deceased. In another case report, more complications. Recently, treatment with nitazoxanide for albendazole only (with less than 50% success rate), but causes improve the cure rate, in comparison to treatment with combination of albendazole and praziquantel may relieve relentless disease. Surgery is often only palliative. Prolonged bony lesions has been reported. However the success of any mode of cysticidal treatment is limited and more effective and tolerable chemotherapy is required.

In conclusion, a high index of suspicion must be held when treating persistent infected non-unions of fractures and multiple cultures and pathology samples must be sent. Extensive debridement must be preformed, with intent, if possible to remove the entire lesion. The fracture must be fixed with a rigid fixation maintaining overall limb alignment, whilst maintain a long term anti-helmintic therapy. Our case demonstrated that this treatment policy achieved a favourable result.

References