Case Report and Review of the Literature

Hydatid disease of bone

M Laubscher MBChB(UFS), Dip PEC(CMSA)
Senior Medical Officer
PS Hardcastle MBChB(US)
Senior Medical Officer
PH Naudé MBChB(UP)
Senior Medical Officer
TN Barrett MBChB(UP), MMed Ortho(UP)
Principal Specialist and Head of Department

Department of Orthopaedics, George Provincial Hospital, George

Reprint requests:
Dr M Laubscher
15 Arundel Street
Rondebosch
7700 Cape Town
Email: maritzl@msn.com
Tel: +27(0)83 384 1191
Fax: 0866 722 198

Introduction

Hydatid disease of bone is uncommon. In endemic areas it should be considered in the differential diagnosis of lytic bone lesions. We present a case of primary hydatid disease of the humerus.

Case report

A 41-year-old gardener was referred to our orthopaedic clinic during January 2007. He presented with the complaint of recent onset of pain in his left arm following minimal trauma. Radiographs revealed a spiral fracture of the left humerus involving the proximal diaphyseal region. Lytic lesions were present around the fracture and it was thought to be a pathological fracture. No significant abnormalities were found during routine blood tests. The white cell and differential counts were normal and the ESR 15 mm/h *(Figure 1)*.

The patient was booked for a biopsy of the lesion. Surgical exposure of the proximal humerus through an antero-lateral approach revealed a multilocular cyst extending from the bone into the adjacent soft tissue. Several daughter cysts were found. The bone was curetted and the wound was washed and closed in layers. Because hydatid disease was suspected samples of the fluid and cysts were sent for histology and microbiology tests. His arm was immobilised in a u-slab.

Microscopy confirmed the diagnosis of hydatid disease. Oral chemotherapy with albendazole 10 mg/kg/day in two divided doses was initiated. The patient recovered uneventfully and no wound or other postoperative problems were detected. A 16-week course of postoperative chemotherapy (three courses of 28 days each, with a 2-week interval between courses) was completed as per guidelines according to the *SAMF*.[1]

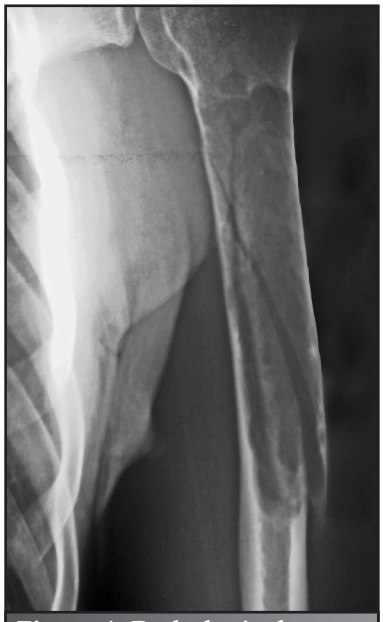

Figure 1: Pathological humerus fracture

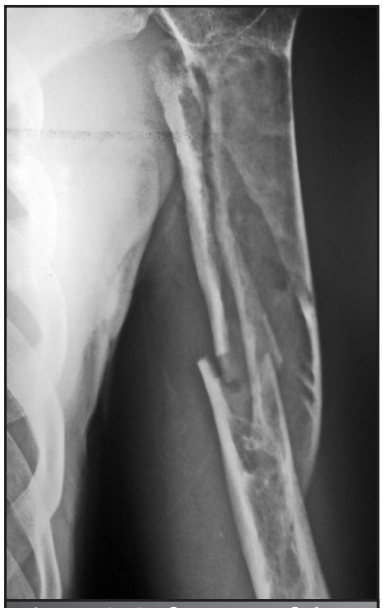

Figure 2: Refracture of the humerus

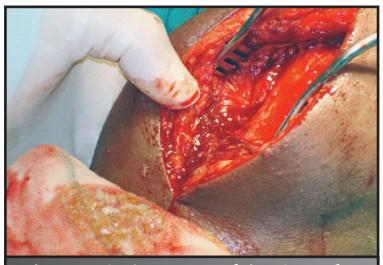

Figure 3: 'Grape skins' and cysts

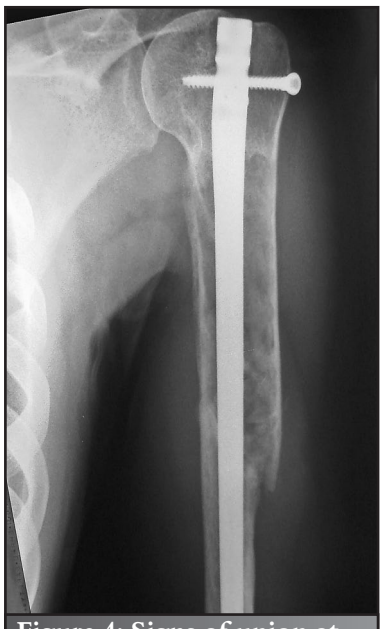

Figure 4: Signs of union at six weeks postoperatively

He was making a good recovery when last seen 7 months postoperatively, with clinical and radiological signs of fracture union. He was then lost to further follow-up.

He presented again one year after his initial fracture with a re-fracture of his left humerus after lifting a heavy object. Radiographs revealed a new fracture with some comminution below the previous fracture site. Marked osteopaenia was evident and it appeared that the lesion now extended from the metaphysis down the diaphysis of the humerus. Routine blood tests, including non-reactive HIV serology, revealed no significant abnormalities. Radiographs of the chest, spine and long bones and an abdominal ultrasound did not reveal any evidence of other sites of hydatid disease *(Figure 2)*.

Surgical exploration of the fracture through an antero-lateral approach to the humerus revealed a multiloculated cystic mass extending from the bone into the soft tissue. The length of humerus was curetted and many 'grape skins' and cysts were removed. After curettage, the medullary cavity and the wound were thoroughly washed with 3% hypertonic saline as scolecidal agent. The wound was packed with swabs soaked in 3% hypertonic saline.

Through a separate deltoid-splitting approach, an interlocking intramedullary humeral nail was inserted. The medullary cavity was reamed to remove any further cysts. Autogenous cancellous bone graft was harvested from the iliac crest and the medullary cavity was packed with the bone graft. Care was taken not to contaminate the proximal entry site of the nail. The wounds were washed and closed in layers. The arm was protected postoperatively in a sling and pendular exercises were initiated *(Figure 3)*.

Legislation in New Zealand making it illegal to feed raw offal to dogs has drastically reduced the incidence of hydatid disease

Oral chemotherapy with albendazole 10 mg/kg/day in two divided doses was prescribed to prevent further recurrence. The patient recovered uneventfully and was discharged postoperatively on day 5. There was no wound or other complications. The patient completed 12 weeks of postoperative chemotherapy with albendazole. He regained full range of motion of his shoulder and elbow joints and returned to work 6 weeks after surgery. He was followed up clinically and radiologically for a further 6 months and there is, at present, no evidence of recurrence. The fracture has united both clinically and radiologically *(Figure 4)*.

Discussion

Incidence

Hydatid disease was known as early as in the time of Hippocrates. It is prevalent in most sheep-raising countries.[2] Hepatic (75%) and pulmonary (15%) involvement is common. Only 10% of lesions occur throughout the rest of the body.[2,3] Skeletal hydatid disease is rare and is found in only 0.5 to 2.5% of cases.[2,4] The spine is the most common skeletal site (35 to 50%),[2,4] followed by the pelvis and hip.[2] Involvement of the humerus has been reported to occur in 2% to 6% of cases of skeletal hydatid disease.[4] Musculoskeletal lesions usually occur in isolation without concomitant hepatic or other organ involvement.[5,6]

Pathophysiology

Hydatid disease (Echinococcosis) is a parasitic disease caused by the larva of the tapeworm *Echinococcus granulosus*, or less commonly *E. multilocularis*.[3,7] The adult tapeworm is found in the intestines of dogs or other canines. Eggs are released in their stools and ingested by the intermediate hosts, usually sheep, where they traverse the intestinal mucosa to enter the portal circulation.[7,8] Canines contract the parasite by eating infected tissue to complete the cycle. Legislation in New Zealand making it illegal to feed raw offal to dogs has drastically reduced the incidence of hydatid disease.[8]

Humans are accidental intermediate hosts.[3] After entering the circulation, the scolex has to pass the filters of the liver and lung before it is carried to bone. This would explain the low incidence of hydatid bone disease.[8] The disease usually starts in the metaphysis of bone before it spreads slowly down the diaphysis.[9,10] Daughter cysts extend into bone, replacing the medulla.[4] Pressure absorption of bone takes place leading to expansion of the lesions.[9] Joint involvement is usually due to secondary extension from adjacent bone, although primary hydatid synovitis has been reported.

Diagnosis

Hydatid bone disease usually remains asymptomatic for a long period.[4,10] It is rarely seen in children, although it is most likely acquired during the early years.[9] The clinical manifestations may take 10 to 20 years to become evident[2,4,9] and normal presentation is in the age group 30 to 50 years.[9] There are no characteristic signs or symptoms. Hydatid bone disease usually presents as a pathological fracture, secondary infection of the cystic mass or with persistent pain.[4] Spinal lesions may present with a myelopathy.

The disease is difficult to diagnose and manage. The diagnosis is usually made at an advanced stage of the disease.[10] Hydatid disease should be included in the differential diagnosis of any expanding osteolytic bony lesion, especially in endemic areas. This will facilitate earlier diagnosis.[2,10,11] The differential diagnosis of skeletal hydatid disease should include the following: pyogenic infections, tuberculosis, fibrous dysplasia, benign cystic conditions, bone tumours, multiple myeloma and metastatic malignancies.[3,10,11,12] The diagnosis is often only made intra-operatively.[8,12] Reports exist of practitioners mistaking the lesion for an abscess and attempting incision and drainage.[13]

Plain radiographs are not diagnostic. Radiological findings range from monolocular to multilocular cysts. Monolocular cysts are rarely observed.[4,10] Multilocular cysts with a honeycomb appearance are most common. Osteolysis is usually seen with expansion of the bone and thinning of the cortex.[2,14] These signs are not specific, but large expanding bone lesions together with soft-tissue calcifications are highly suggestive of hydatid disease.[12,14]

CT has better bone resolution, and usually shows irregular erosions of bone.[12] It is also more accurate in delineating the area of destruction and determining extra-osseous spread.[2] MRI with its superior soft-tissue resolution is useful to determine extra-osseous extension of the disease.[12] Technetium bone scanning is not reported to be helpful in diagnosing the disease.[3]

Serological tests may be useful in suspected cases. Detection of antibodies is more sensitive than the detection of antigens.[2] The sensitivity of ELISA, indirect haemagglutination and the complement fixation test drops to anything between 25 to 50% in bone disease.[12] The Casoni intradermal test is non-specific.[7] Serodiagnosis may be useful for follow-up after surgical or pharmacological treatment.[2] Eosinophilia is seen in only 25 to 30% of cases.[4,7] FNA is not advocated due to the risk of cyst rupture which can lead to dissemination of the disease and anaphylactic reactions.[3,12] The definitive diagnosis can normally be made by histopathological examination.[4] The diagnosis of recurrent disease should not solely rely on imaging, but should be based on a combination of symptoms and serological tests.[12]

Treatment

The treatment of hydatid bone disease includes a combination of surgery and systemic and local chemotherapeutic agents.[9] Treatment is complicated by the fact that it is a very uncommon condition and the diagnosis is often made late or only at or after surgery. It has been suggested that once the disease has developed in its reticular form in bone, it is untreatable.[9] Skilful neglect seems to be selected as a last resort by some physicians.[2]

Most authors advise radical surgical excision with wide safety margins as the only definitive treatment of the condition.[3,10,15] This may not be possible due to the site of the lesion. It has not been fully established how hydatid disease in bone spreads and therefore it is difficult to establish a safe surgical margin. In 1987, Szypryt *et al* reported success with the use of less radical surgery with combination chemotherapy in five patients.[16] The lesions were curetted and bone was grafted in combination with chemotherapy with albendazole.[10]

The treatment of pathological fractures should follow normal protocols. It is suggested that large defects should be filled with autogenous bone graft.[4] Polymethylmethacrylate has also been used.[14]

The necrotising effect of the heat of the polymerising cement may kill the daughter cells.[14] Wide resection followed by replacement with a custom-made megaprosthesis has also been reported successfully.[11] This is not readily available in all centres and potential recurrence in the same place would be disastrous.

The combination of systemic chemotherapy and surgery improves the probability of cure.[3,8,16] Isolated reports of successful treatment with chemotherapy alone exist, although this is extremely rare.[8,12] High doses of albendazole (10 mg/kg/day) and mebendazole (40 mg/kg/day) are recommended.[2] These high dosages cause side-effects, with albendazole being less toxic.[7] The reported duration of treatment varies. A minimum of six courses of albendazole (each course consisting of 28 days of treatment), appears to be needed.[2] At least one course should be given pre-operatively.[2] There are reported cases of treatment with chemotherapy for more than two years.[7]

The combination of systemic chemotherapy and surgery improves the probability of cure

Praziquantil has been proven to be effective in animal trials, although no evidence of efficiency in humans exists. It is suggested that the actions of albendazole and praziquantil in humans may well be synergistic.[2]

Local chemotherapeutic agents have been added to the treatment of hydatid bone disease intra-operatively in an attempt to improve local eradication of the disease. The most commonly used agents include the following: 3% hypertonic saline, 10% formalin, hydrogen peroxide, chlorhexidine, 80% alcohol, povidone-iodine-alcohol solution or 0.5% silver nitrate.[9,10,12] These agents destroy daughter cysts, but may not be completely effective and therefore do not necessarily prevent local recurrence.[9,14] Radiotherapy has been reported to be ineffective in the treatment of hydatid disease of bone.[10] It was thought that secondary staphylococcal infection may play a role in killing hydatid parasites, although poor evidence exists.[9]

Complications

Potential complications include deformity, pathological fracture, secondary bacterial infection and the formation of fistulas.[7,10] Pathological fractures tend to produce a non-union.[14] Pressure on adjacent tissues may cause pain and neurological deficit (in the case of spinal involvement).[2] Rupture and spillage of cyst content is known to provoke hypersensitivity.[12] Death has even been reported due to anaphylactic shock resulting from spillage of a ruptured cyst during biopsy.[3]

Prognosis

The prognosis of hydatid bone disease is poor. Local recurrence occurs in approximately 40% of cases, despite aggressive treatment.[12] Recurrences may occur 2 to 28 months following the initial diagnosis or procedure.[12] Wide local excision is not always possible and intra-operative seeding is almost impossible to prevent.[12] The mortality rate varies from 0% to 3% and is mostly associated with spinal hydatid disease due to progressive myelopathy.[12] Involvement of other organs should be excluded, although musculoskeletal hydatid disease usually occurs in isolation.[2,5,6] Advances in imaging and chemotherapy have improved the prognosis.

Summary

Hydatid disease should be considered in the differential diagnosis of lytic bone lesions in endemic areas like South Africa. Musculoskeletal hydatid disease usually occurs in the absence of other systemic diseases[2,5,6] and can prove extremely difficult to treat. Numerous complications can occur. Surgery with adjuvant local and systemic chemotherapy would appear to offer the best treatment option. Long-term survival is possible, but the disease is not easy to eradicate and may well be impossible to cure.[10]

This article has been submitted to an ethical committee and is awaiting approval. The contents of this article is the sole work of the authors.

No benefits of any form have been derived from any commercial party related directly or indirectly to the subject of this article.

References

1. Gibbon JG *et al. South African medicines formulary*. (6th edition). Cape Town: SAMA Health and Medical Publishing Group. 2001;468-72.
2. Farzan M, Mortazavi SMJ, Motamedi M. Hydatid disease of bone. *Acta Medica Iranica* 2006;**44**(6):361-4.
3. Omidi Kashani F, Hagi Z, Mazloumi M. Unusual presentation of iliac bone hydatid cyst. *Iran Journal of Medical Science* 2007;**32**(2):121-3.
4. Fanian H, Karimian Marnani M. A case report of hydatid disease in long bone. *Journal of Research in Medical Sciences* 2005;**10**(2):101-4.
5. Merkle EM, Schulte M, Vogel J *et al.* Musculoskeletal involvement in cystic echinococcosis: report of 8 cases and review of the literature. *American Journal of Roentgenology* 1997; **168**(6):1531-4.

References continued on page 70.

Hydatid disease of bone

References continued from page 47.

6. Garcia-Alvarez F, Torcal J, Salinas JC, Navarro A, Garcia-Alvarez I, Navarro-Zorraquino M *et al.* Musculoskeletal hydatid disease: a report of 13 cases. *Acta Orthopaedica Scandinavia* 2002; **73**(2):227-31.
7. Belzunegui J, Maíz O, López L, Plazaola I, González C, Figueroa M. Hydatid disease of bone with adjacent joint involvement: A radiological follow-up of 12 years. *British Journal of Rheumatology* 1997;**36**(1):133-5.
8. Alldred AJ, Nisbet NW. Hydatid disease of bone in Australasia. *Journal of Bone and Joint Surgery* 1964;**46**B(2):260-7.
9. Hooper J, McLean I. Hydatid disease of the femur: Report of a case. *Journal of Bone and Joint Surgery* 1977;**59**A(7):974-6.
10. Booz MK. The management of hydatid disease of bone and joint. *Journal of Bone and Joint Surgery* 1972;**54**B(4):698-709.
11. Natarajan MV, Kumar AK, Sivaseelam A, Iyakutty P, Raja M, Rajagopal TS. Using a custom mega prosthesis to treat hydatidosis of bone: a report of 3 cases. *Journal of Orthopaedic Surgery* 2002;**10**(2):203-5.
12. Necmettin Pamir M, Özduman K, Elmaci I. Spinal hydatid disease. *Spinal Cord* 2002;**40**:153-60.
13. Ranga Chari P. Hydatid disease of scapula and upper third of humerus treated by en bloc excision and fibular bone grafting. *Indian Journal of Orthopaedics* 2007;**41**(3):241-4.
14. Yildiz Y, Bayrakci K, Altay M, Saglik Y. The use of polymethylmethacrylate in the management of hydatid disease of bone. *Journal of Bone and Joint Surgery* 2001;**83**B:1005-8.
15. Kalinova K, Proichev V, Stefanova P, Tokmakova K, Poriazova E. Hydatid bone disease: a case report and review of the literature. *Journal of Orthopaedic Surgery* 2005;**13**(3):323-5.
16. Szypryt EP, Morris DL, Mulholland RC. Combined chemotherapy and surgery for hydatid bone disease. *Journal of Bone and Joint Surgery* 1987;**69**B:141-4.