Primary Hydatid Disease of the Humerus

Dear Editor,

Introduction

Hydatid disease is a parasitic tapeworm infection caused by *Echinococcus* species. *E.granulosus* and *E.alveolaris* are the most common causes of hydatid disease in humans. Hydatidosis is endemic in the sheep-raising districts of Australia, South America, Central Asia, the Middle East and the Mediterranean countries. Osseous involvement is rare and bone lesions are reported in only 0.5% to 4.0% of the cases. Isolated primary hydatid disease of the bone without hepatic or pulmonary involvement is extremely rare, as the the parasite has to cross pulmonary and hepatic barriers before reaching the bone.[1] Moreover, the humerus is an unexpected site for osseous involvement.[2-4] Due to its rarity, it often presents a clinical and radiological diagnostic challenge. Treatment of osseous lesions is also difficult because of the invasive nature of bony involvement and the spillage of fluid with subsequent seeding.[1,5]

Case Report

A 56-year-old man was admitted to our hospital with a sudden pain in his left shoulder after he bumped into a door. On physical examination, there was swelling and tenderness over his shoulder and upper arm with limited motion. His past medical history was unremarkable.

Plain film of the left humerus demonstrated numerous radiolucent areas at humeral metaphysis extending to the diaphysis with cortical thinning. A non-displaced pathologic fracture at the proximal diaphysis was visible. No soft tissue calcification was noticed. Magnetic resonance imaging (MRI) revealed a multi-cystic lesion at the humeral metaphysis with extension to the diaphysis down to the supracondylar region. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images and showed heterogenous enhancement after intravenous administration of gadolinium contrast. Diffuse cortical thinning and breakage was noticed throughout the diaphysis without soft tissue involvement (Fig. 1). Scintigraphy showed no abnormal activity in the whole skeleton. With the assumption of a bone metastasis of a solid organ tumour, a CT scan of the whole abdomen, thorax and cranium was performed, but no other lesion was detected.

Based on all these clinical and radiological data, the mass was suspected to be a primary benign bone tumour or an infectious process. Curettage, grafting and fracture fixation were planned. During the operation, a massive multicystic lesion in the metaphyseal area that extended to the diaphysis and the supracondylar region of the humerus containing soft white tissue was found and removed. Extensive curettage and irrigation followed by filling the cavity with massive allograft chips was performed. Thereafter, plate and screw fixation was applied to the fracture. Histopathological examination of the biopsy material revealed hydatid disease. This was an unexpected diagnosis. Albendazole (10 mg/kg) was administered and continued for 3 months with monitoring of the hepatic enzymes. One year after surgery there was instability due to loosening and progress of the disease. Thereafter, additional plate and screw fixation with further curettage and grafting was performed.

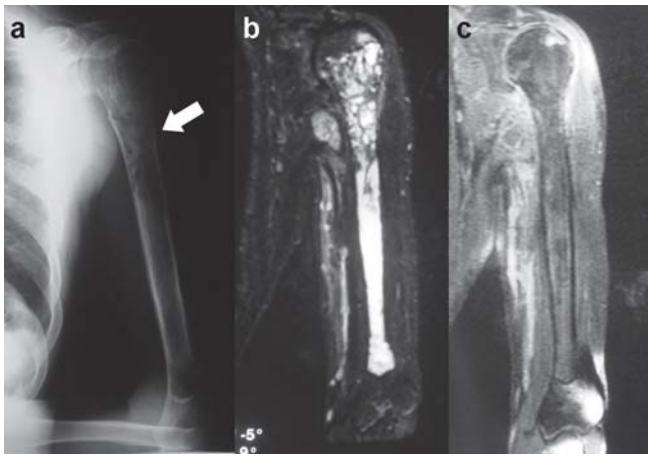

Fig. 1. (a) Direct X-ray of the humerus, white arrow shows the pathologic fracture line. (b) Coronal T2 weighted sections of the lesion. (c) Coronal T1 weighted sections of the lesion.

Discussion

Hydatid disease in bones occurs mostly in vascularised areas. The vertebra, long-bone epiphyses, ilium, skull and ribs are most frequently affected in descending order. The vertebral column is involved in approximately half of osseous patients due to porto-vertebral shunting.[5] The humerus is an extremely rare site for primary hydatid disease without hepatic and pulmonary involvement.[2-4]

Radiological features of the osseous hydatid disease are well-defined. It produces multiloculated osteolytic lesion with cortical thinning and possible bone expansion. Marginal sclerosis, periosteal reaction and soft tissue invasion are uncommon. Periosteal reaction can be observed in case of infection. These signs are not specific, but large lesions with adjacent soft tissue calcifications are highly suggestive of hydatid disease.[5] Prior to MRI, computed tomographic (CT) scanning played a major role in the

evaluation of osseous involvement. Calcified or uncalcified cysts may be seen with expansion and cortical thinning. MRI imaging is the most comprehensive of all imaging modalities. The characteristic feature of this disease is the presence of multiple cysts within a cavity, which are hypointense on T1-weighted images and hyperintense on T2-weighted images. The MRI should be the choice of evaluation in order to appreciate the extent of disease and surgical planning.[2]

The radiological appearance of the hydatid disease in bone is not specific and mimics tumours and other inflammatory conditions. Solitary lesions in bone can be mistaken for a plasmacytoma, a simple bone cyst or a brown tumour of hyperparathyroidism. A botryoid configuration of the cysts could resemble a chondromyxoid fibroma or skeletal metastasis. A soft tissue component adjacent to a lytic bone lesion would raise the differential of a chondrosarcoma, osteomyelitis (pyogenic/tuberculous) or a giant cell tumour or an aneurysmal bone cyst. An infiltrative pattern of lucencies simulates multiple myeloma. Fibrous dysplasia can closely resemble long-standing cases of hydatidosis in which the marrow cavity is extensively invaded by the parasite as in the presented case. A conclusive diagnosis of hydatidosis could be reached in only half of the cases preoperatively.[5]

Serologic tests are valuable when they are positive but they are usually false negative for osseous hydatidosis. Complete reliance on the serology is not recommended for exact diagnosis.[1] As we determined the diagnosis from histopathological examination, serologic tests were unnecessary.

Complete cure of the osseous involvement is a therapeutic challenge. The disease generally is not curable and recurrence is likely. Surgery is the treatment of choice in osseous echinococcosis, although it can only eliminate the macroscopic cysts and local recurrence is reported in half of patients. Treatment should include a combination of surgery and chemotherapy with albendazole. Osseous hydatid disease behaves like a locally aggressive relapsing bone tumour.[1] Skeletal tumour treatment strategies such as total excision with either biologic or prosthetic reconstruction are good alternatives in long bone involvement.[5] However, when the lesion diffused rather than situated on a segment, it is impossible to preserve the extremity.

The diagnosis of primary bony hydatidosis is difficult. It may be easily overlooked and usually requires a high index of suspicion. In endemic areas, hydatid disease should be considered in the differential diagnosis of any cystic lesions of bones. Preoperative diagnosis is essential for correct surgical planning and prevention of recurrence.

REFERENCES

1. Arazi M, Erikoglu M, Odev K, Memik R, Ozdemir M. Primary echinococcus infestation of the bone and muscles. Clin Orthop Relat Res 2005;432:234-41.
2. Markonis A, Tavernaraki A, Papaevangelou M. Humeral hydatid cyst complicated with extraosseous involvement: a case of unusual location of echinococcosis. Eur J Radiol 2001;37:130-3.
3. Aslan B, Aslan G, Ozardali I. Multilocular echinococcosis in humerus: a case report. J Egypt Soc Parasitol 2001;31:961-2.
4. Pintilie DC, Panoza G, Hatmanu D, Fahrer M. Echinococcosis of the humerus. Treatment by resection and bone-grafting: A case report. J Bone Joint Surg Am 1966;48:957-61.
5. Morris BS, Madiwale CV, Garg A, Chavhan GB. Hydatid disease of bone: a mimic of other skeletal pathologies. Australas Radiol 2002;46: 431-4.

Hamdi Ozkan,[1]*MD*, Yunus Dogramaci,[2]*MD*, Ozkan Kose,[3]*MD*, Erdinc Esen,[2]*MD*, Hakan Erdem,[4]*MD*, Mahmut Komurcu,[3]*MD*

[1] Department of Orthopaedics and Traumatology, Ataturk Education and Research Hospital, Ankara, Turkey
[2] Department of Orthopaedics and Traumatology, Gazi University, Medical Faculty, Ankara, Turkey
[3] Department of Orthopaedics and Traumatology, Gulhane Military Medical Academy, Ankara, Turkey
[4] Department of Infectious Diseases and Clinical Microbiology, Gulhane Military Medical Academy, Ankara, Turkey

Address for Correspondence: Dr Ozkan Kose, Muminderesi cd. Belediye Bloklari, 3. Blok Daire: 10, Sahrayicedid, Kadikoy, Istanbul, Turkey.
Email: drozkankose@hotmail.com