REVIEW ARTICLE

Spinal hydatidosis

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Introduction

Hydatid disease is caused by the larval forms of the cestode worm, Echinococcus granulosus. Only two species, Echinococcus granulosus and Echinococcus multilocularis are known to affect man. E. granulosus is more benign and is characterised by cyst formation, while the multilocularis type, which manifests as the rare alveolar form, is not encapsulated and presents as a porous and ramifying necrotic mass. The prognosis for the alveolar form is extremely poor with an almost 100% mortality rate. Echinococcosis rarely involves the skeleton but mainly affects the liver and the lungs. The prognosis for neurologic recovery in spinal hydatid disease is poor and posterior surgical decompression is associated with a high recurrence rate. Vertebrectomy and excision of the posterior elements when indicated and antihelmintic drugs are useful to control the disease and prevent recurrence. This review highlights the pathology, clinical presentation and treatment.

Epidemiology

Hydatid disease in man is universal but is particularly common in areas where sheep are raised, notably in North Africa, Greece, Yugoslavia, South Africa, East Africa, South America and north western Canada.[1-10] The infestation may be transmitted from endemic areas by tourists.[7-9] The incidence of echinococcosis infestation is increasing and, apart from endemic regions, the infestation has been reported in areas previously free of it.[2,9,11,12]
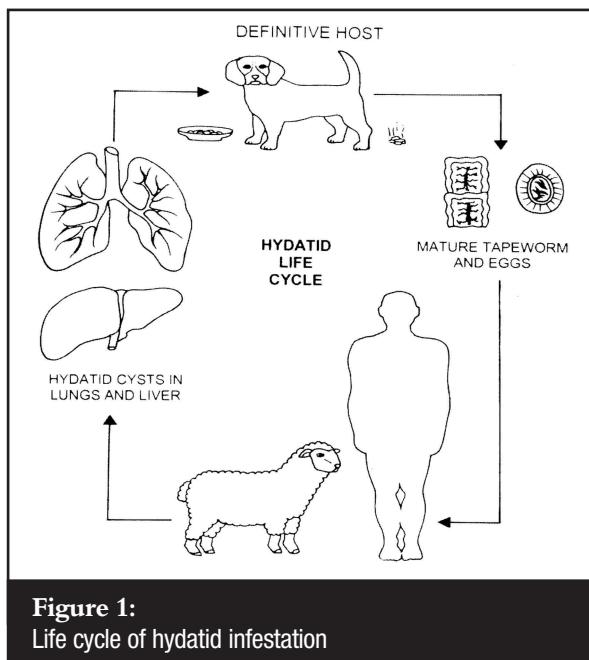
Life cycle

The infestation is caused by ingestion of the ova of Taenia Echinococcus *(Figure 1)*. The definitive host of the adult worm is usually the domestic dog but other canines including wolves, foxes and jackals are equally important hosts. The host is infested by eating the viscera of an intermediary host harbouring the hydatid cysts. The sheep is the common intermediary host for the larval worm, but cattle, hogs and in north-western North America, caribou and moose may also be infected. The scolices within the cysts develop into mature worms in the intestines of the definitive host and the ova are then excreted in the faeces.

Man acquires the infection by ingestion of the ova which are usually found in the fur of infected dogs.[5] The chitinous coat of the ova is dissolved in the intestine of the intermediate host. The embryos which are liberated pass through the intestinal wall into the portal venous system and develop within weeks into larvae which then produce cysts. About 75% of the larvae are trapped in the liver, 15% in the lungs and the remaining 10% are disseminated elsewhere in the body.[12-14]

Pathology

Echinococcosis manifests itself differently in bone than in the liver or the lungs due to the mechanical resistance that bone offers to the growth of hydatid cysts.[7,8] Osseous involvement occurs in 0.5% to 2% of all cases of hydatidosis.[15-21] The bones most often involved are the vertebrae (44%), long bones (38%), ilium (16%), skull (4%), ribs (3%), scapula (1.5%) and the sternum (0.8%).[22-27] When embryos are deposited in bone, destruction occurs by mechanical pressure without an inflammatory reaction.

Figure 1:
Life cycle of hydatid infestation
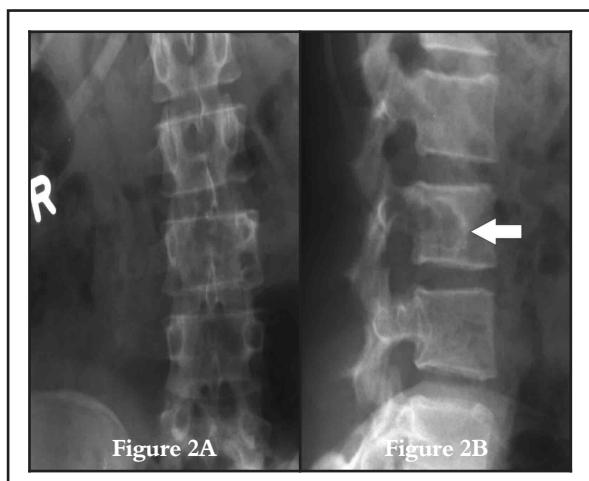
Figure 2A   Figure 2B

Figure 2:
AP/LAT radiograph show lytic lesion with sclerosis (arrow) and involvement of the posterior elements

There is no pericyst formation as in soft tissues (liver and lung), and the parasite expands by exogenous proliferation in an irregular branching fashion. The expansion occurs along the lines of least resistance because of the small size of the bony canals and the resistant nature of their walls. The parasite grows in all directions invading the vertebral body and the posterior elements of the vertebra and resembles a slow-growing tumour. In the thoracic spine, rib involvement is common.[28,29]

> The vertebral column is the most common
> site of skeletal involvement in hydatid disease

Clinical features

The vertebral column is the most common site of skeletal involvement in hydatid disease. Vertebral hydatidosis is a relatively silent, slowly progressive disease with a latent period of many years.[1,18,30] The clinical manifestations are variable depending on the localisation of the hydatid cyst. The infestation usually affects adults and is rare in children. Gharbi et al[31] reported only one case in a child and Slim et al[32] found no cases of osseous involvement among 34 children with hydatid disease of other organs. Pain, which is an important presenting symptom,[2,28,29] begins insidiously and becomes progressive. There are no accompanying features of fever or weight loss. Iliac fossa pain due to lumbar vertebral disease may simulate an abdominal syndrome.[30] Thoracic spine involvement may manifest with pain radiating into shoulders and scapula. In endemic areas, hydatid disease is a common cause of progressive spinal cord compression which may progress to paraplegia.[16,18,33] Cysts invade the spinal canal and cause spinal cord or cauda equina compression leading to motor, sensory and sphincter disturbance.[34-36] Radicular pain due to rib involvement may precede paraparesis in lesions affecting the thoracic spine. Cysts in the lumbar spine may cause nerve root compression and mimic sciatica. The clinical examination may reveal a soft tissue mass in the iliac fossa or over the lateral chest wall. Kyphosis or kyphoscoliosis in the thoracic region has been reported following extensive disease.[17,21,35] Hydatid disease of the spine should be considered in the differential diagnosis in non-endemic areas.

Laboratory findings

The erythrocyte sedimentation rate (ESR) and eosinophil counts have the little value. Detection of circulating antigen (CAG) and circulating immune complexes (CIC) have been reported to be helpful in monitoring the disease.[23,37]

Imaging

Identification of hydatid disease on plain radiographs is difficult.[26,34] However, there are some radiologic features that may indicate the presence of spinal echinococcosis.[18] The thoraco-lumbar spine is the commonest area of involvement. Anteroposterior and lateral radiographs may reveal asymmetric osteolysis of the vertebral body and the neural arch *(Figures 2a,b)*. The lesion in the vertebral body and the posterior arch is translucent, circular or oval and may result in expansion and thinning of the cortices. Early in the disease the osteolysis is well defined with a sclerotic margin.[37] Usually there is no reactive new bone formation. Narrowing of the disc space and collapse of the vertebral body are late features. In the thoracic and lumbar spine, paravertebral soft tissue swelling may be a feature. The disc is reported to be relatively resistant to invasion and extension to adjacent vertebrae occurs beneath the anterior longitudinal ligament.[10,36,38]
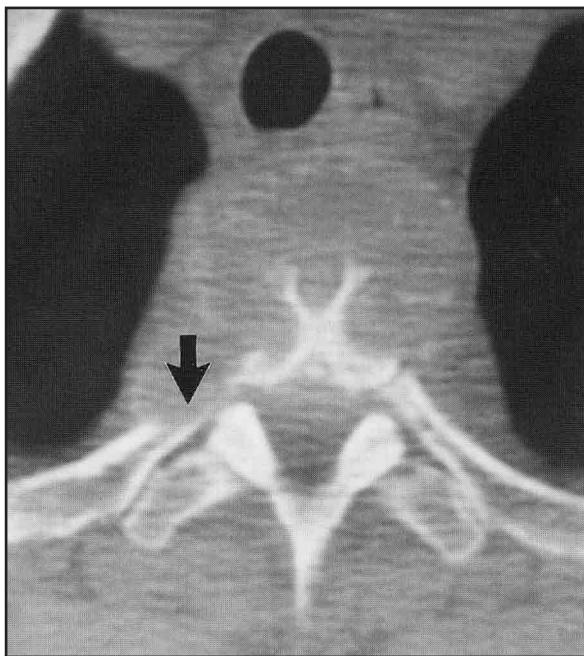
Figure 3:
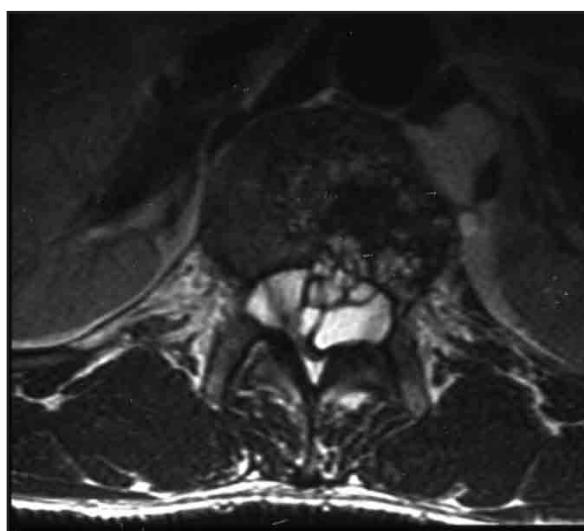Arrow demonstrates rib involvement (arrow) and the vertebral body with a paraspinal mass

Figure 4:
Axial CT showing paraspinal extension (arrow) with asymmetric involvement of vertebral body

Progression of the lesion to the ribs and iliac bone is characteristic of spinal echinococcosis *(Figure 3)*. All patients with thoracic disease had rib involvement in the series reported by Karray *et al.*[28]

Ultrasound is a useful investigation to evaluate an abdominal mass in symptomatic patients and to detect liver and paraspinal involvement. Computerised tomography (CT) scan depicts the degree of vertebral destruction and may demonstrate intra- and paraspinal involvement *(Figure 4)*.[28] MRI is superior to CT in highlighting the osseous, soft tissue extent and the precise localisation of cord compression *(Figure 5)*. The MRI scan characteristically shows an image resembling a bunch of grapes. The cyst walls are thin and regular with no septations. The cerebrospinal fluid and cyst content essentially have the same signal intensities on all sequences. The presence of a markedly hypointense cyst wall on T1- and T2-weighted images and the absence of wall enhancement with gadolinium are characteristic of hydatid disease. The MRI has been utilised as the study of choice for diagnosis and analysis at follow-up. MRI may be helpful in assessing residual cysts and in detecting recurrence at an earlier stage.[25,28,37]

Figure 5:
Axial CT demonstrating hydatid cyst with marked cord compression

Differential diagnosis

The differential diagnosis includes tuberculosis, metastatic disease, plasmacytoma, haemangioma and giant cell tumour.[34] In tuberculosis there is always some bony reaction, early disc space narrowing, or destruction of the adjacent disc but it rarely involves the posterior elements or a contiguous rib.

Natural history

The prognosis of vertebral hydatid disease is poor especially when neurological symptoms and signs are present.[28] Vertebral involvement is life-threatening with a reported mortality in excess of 50%. In Britain the average length of survival after the onset of symptoms was five years and the average age at death was 41 years.[38]
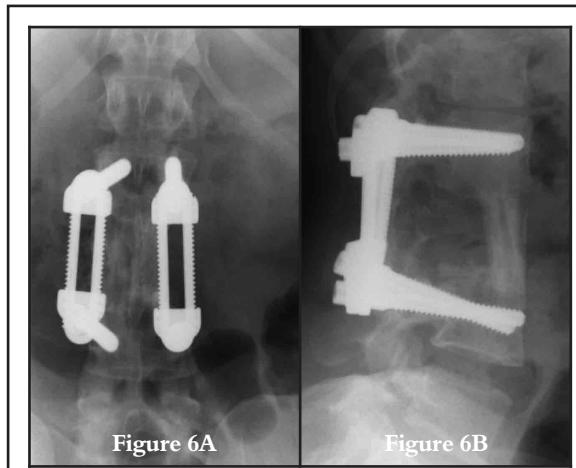
Figure 6:
AP/LAT view at 16 year follow-up without recurrence following anterior and posterior surgery

Treatment

In the past the treatment of osseous hydatid disease with neurological deficit has been entirely surgical which entailed removal of the cyst and surrounding bone through an extensive laminectomy.[5,26,28] The surgical goals were rarely achieved in this relentless disease and frequently a second-look procedure was undertaken to detect early recurrence following laminectomy. In the long term laminectomy alone was unsatisfactory because of progressive deformity resulting from instability and recurrence following inadequate decompression. The long-term outcomes of spinal hydatid disease after posterior decompression were reported by Apt et al[18] who performed 69 operations in 26 patients for recurrence during 15 years. They concluded that posterior spinal cord decompression was incomplete because of the failure to remove all cysts. Turtas et al[39] reported a recurrence rate of 50% one to six years after posterior decompression and recommended extensive resection of the vertebral bodies and fusion to control hydatidosis. They reported that complete recovery from paraplegia was unusual and the prognosis for recovery was influenced strongly by the neurologic status at the time of surgery. Although posterior decompression and instrumentation in hydatid disease has been shown to correct and prevent kyphosis, it has not consistently improved the neurological status.[28]

Treatment of hydatid disease of the spine is a combination of early surgery and antihelmintic drugs[37,40-42] Preoperative evaluation includes plain X-rays, CT scan, MRI and ultrasound. These investigations are helpful to delineate the lesion and in the surgical planning. The liver and the lungs, which are the primary target organs, should be evaluated by MRI or CT scan.[37] This is of therapeutic importance because in the absence of hepatic or pulmonary disease a meticulous surgical excision of the osseous lesion can be considered curative.

Ideally treatment entails as wide a resection as possible to provide complete removal of the lesion. Because of the difficulty of total excision of one or more affected vertebrae, and the surrounding soft tissue, partial resection is often necessary. A combined approach consisting of posterior decompression, instrumentation, corpectomy, cyst removal and anterior interbody fusion *(Fgure 6)* is recommended.[37] In patients with neurologic deficit remission is obtained in almost all cases for a period of 15 months (av) following laminectomy and five years or more in patients operated by the combined approach.[28,37]

Duran et al[15] and Booz[43] recommended the use of chemical sterilisation of the scolices with hypertonic saline, formalin or 0.5% silver nitrate during surgical removal of the cysts. The scolecidal agents may be curative but do not destroy all cysts and therefore recurrence and dissemination may occur.

There are few reports on the duration and dosage of chemotherapy for osseous lesions.[13,14,44,45] Albendazole reduces the viability of protoscolices and cysts and is also active against the larval cestodes. Albendazole sulfoxide is better absorbed with higher levels of the active metabolite in the cysts compared with the other benzimidazoles.[46] Six courses of albendazole have been recommended for Cag levels to become negative which is an indicator of viability and biological activity of the parasite.[23,47] Following surgery we use albendazole (10 mg/kg/day) for six cycles of 25 days each. It is essential to monitor the liver function during treatment for hepatotoxicity. We concur with others that antihelmintic therapy improves the prognosis of vertebral hydatidosis when combined with surgery.[37]

Conclusion

Prevention of echinococcosis by eradication of the parasite from the pool of primary hosts as was undertaken in Iceland and New Zealand is the only way to avoid infestations. The prognosis for neurological recovery and a disease-free interval of greater than a decade is good following early diagnosis, radical excision of the diseased segment and antihelmintic drugs.

The content 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.

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