Primary Spinal Alveolar Hydatid Disease: A Case Series with Review of Literature

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ABSTRACT

AIM: The aim of this case series is to share our experiences of the pitfalls and challenges in the diagnosis and management of 3 cases of primary spinal alveolar hydatid disease over 5 years and to provide a perspective through review of the literature.

MATERIAL AND METHODS: The patients were evaluated clinically and investigated using the relevant imaging modalities. Surgeries by posterior approach were performed with removal of the entire cyst with through debridement along with neural decompression in all. The instability of spine was taken care with instrumentation. Diagnosis was confirmed by histopathologic examination. All patients received long-term antihelminthic therapy with 15mg/kg per day of Albendazole in monthly cycles.

RESULTS: After surgery, all patients improved. However, over time, recurrence was observed. Two patients had complete neurological recovery at regular follow-up, one patient lost to follow-up for about 18 months. Later on she presented with recurrence of disease and subsequently died.

CONCLUSIONS: Primary alveolar spinal hydatid disease is a very rare condition. It is difficult not only to diagnose but to treat as well. Spinal instability should be kept in mind. Regular follow-up is must to decrease morbidity and mortality by detecting recurrence as the rate is very high.

KEY WORDS: Alveolar hydatid, hydatid cyst, spinal

INTRODUCTION

Hydatid disease is a parasitic disease caused by infection with tiny tapeworms of the genus Echinococcus granulosus and multicularis. Echinococcosis is classified as either cystic Echinococcosis or alveolar Echinococcosis that is endemic to the temperate climate. Canines are the primary host. The life cycle of E. granulosus may also involve sheep, cattle, goats, and humans. This infection is transmitted orally via eggs shed in the faeces of infected animals. Hydatid disease is not uncommon in areas where cattle rearing are a common occupation. Churrier was the first person to describe spinal hydatid disease in 1807. Reydellet was credited for the first surgery in spinal hydatid in 1819 (23).

The exact incidence of spinal hydatid is not known due to its rarity but it is about 1%. The incidence of spinal alveolar hydatid disease is even less. Here we are referring three cases of alveolar spinal hydatid disease operated by us.

PATIENTS AND METHODS

We encountered three patients in last five years. All were suffering from back pain not relieved by medication. The average duration of back pain was two years. Out of these three patients one was male (case 2) and two were female (case 1 and 3). All had signs of progressive myelopathy with ASIA (American Spine Injury Association) score of A at the time of presentation to our institute with bladder involvement. One patient (case 2) was operated for some cystic lesion in the right armpit three years back but the histopathology record was not available. Each patient was subjected to various investigations as mentioned below and diagnosed as having alveolar hydatid cyst of thoracic spine (Table 1).
**Investigations**

All the patients were subjected to magnetic resonance imaging (MRI) of thoracic region with screening of the rest of the spine (Figures 1A,B; 2A,B; 6) which showed multiple cystic swellings in extraspinal as well as intraspinal extradural spaces compressing the spinal cord. The cysts were hypointense in $T_1$ images while $T_2$ images shows hyperintensity. Apart from this, all patients were subjected for routine blood investigation, CT scan brain, chest, abdomen and pelvis as screening to rule out other cysts (Figures 3A-C; 5). Antibodies for echinococcus antigen were found positive in all. Histopathology confirmed the diagnosis as alveolar hydatid cyst in surgical specimens.

**Surgical management & technique**

All three patients were subjected to surgery by the posterior approach. Laminectomy with removal of the entire cyst with thorough debridement along with neural decompression was performed (Figure 1C; 4, 7). The instability caused during the procedure was taken care of with pedicle screw fixation at two levels above and below the diseased segments (Figure 3D). Normal surrounding tissue was covered by cotton sponges wet with 3% normal saline to avoid fluid contacts in case of accidental rupture of cysts. Post operatively case 2 developed wound infection that healed by debridement and frequent dressing in the same hospital admission (Table 2).

**Medical management**

All the three patients were given Albendazole 15 mg/kg daily in divided dosages for several one-month cycles separated by two weeks of drug-free intervals. The drug was started preoperatively as soon as the condition diagnosed. All the patients were subjected to a complete blood count as well as liver enzyme titer measurement before starting the drug and monthly thereafter.

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Figure 1: Case 1. A) MRI axial sections showing hydatid cysts in the intraspinal and paravertebral regions. B) MRI coronal sections showing paravertebral extension of the cysts. C) Postoperative specimen of multiple hydatid cysts.

Figure 2: Case 2. A) MRI sagittal section showing hydatid cysts in the intraspinal and paravertebral region with vertebral body destruction. B) MRI axial section showing paravertebral extension of the cysts.
Figure 3: Case 2. A) CT scan thorax coronal and 3D reconstruction showing right side paravertebral and intrathoracic extension of multiple alveolar hydatid cysts and 9th rib destruction, B) CT thorax axial cuts, C) CT thorax coronal section, D) postoperative dorsal spine skiagram anteroposterior & lateral view showing pedicle screw & rods for stabilization.

Figure 4: Case 2. A-C) intraoperative photographs showing multiple spinal hydatid. D) postoperative specimen of multiple spinal hydatid cysts.
RESULTS

Case No. 1: D12-L2 laminectomy with exploration with removal of 50 cysts with spinal instrumentation between D10, 11 and L3,4 was performed. The patient was discharged with MRC grade 4 power and Nurick grade 4 grading. Patient was lost to follow-up for about 18 months. Later on she presented with recurrence of disease and subsequently succumbed to death.

Case No. 2: D10 laminectomy with exploration with removal of multiple cysts with spinal instrumentation between D8, 9 and D11,12 was performed for this patient. Patient developed wound infection that healed in the same hospital admission. Power was MRC grade 4 and Nurick grading was 4 at the time of discharge. Patient improved to Nurick grade 0 on regular follow up with no recurrence.

Case No. 3: D 6 laminectomy with exploration with removal of multiple cysts with spinal instrumentation between D4, 5 and D8, 9 was performed for this patient. Power was MRC grade 4 at the time of discharge and Nurick grading was 4. Patient improved to Nurick grade 3 on regular follow up with no recurrence (Table 2).

DISCUSSION

Hydatid – means ‘watery cyst’ (Greek). Hydatid disease is a parasitic disease caused by infection with tiny tapeworms of the genus Echinococcus. Echinococcosis is classified as either cystic Echinococcosis or alveolar Echinococcosis (also known as coastal Echinococcus). Cystic Echinococcosis (CE), also known as hydatid disease, is caused by infection with the larval stage of Echinococcus granulosus.

Alveolar Echinococcosis (AE) disease is caused by infection with the larval stage of Echinococcus multilocularis, a 1-4 millimeter long tapeworm found in foxes, cats, and dogs (definitive hosts). Small rodents are intermediate hosts for E. multilocularis. Although cases of AE in animals in endemic areas are relatively common but human cases are rare. AE poses a much greater health threat to people than CE, causing parasitic tumors that can form in the liver, lungs, brain, and other organs. If left untreated, AE can be fatal.

In the cycle of hydatid disease, herbivore usually harbors the larvae, mostly in the viscera, and a carnivore becomes infected with the adult tapeworm by eating raw viscera. In turn, the herbivore intermediate host acquires a
larval infection by ingesting dog tapeworm eggs. Humans acquire the infection when they become intermediate hosts of these tapeworms by accidental ingestion of the eggs of Echinococcus species. After entering the body, the eggs transform into cysts that grow in the liver, lungs, heart, and CNS. In the latter, cysts may also result from metastatic dissemination of a visceral cyst.

Although rare in developed countries, the disease continues to be a significant health problem in endemic areas (1, 6, 35). The disease is especially prevalent in areas where livestock is raised in association with dogs: Mediterranean, eastern European, east African, central Asian and South American countries, Australia and New Zealand. Sapkas et al (39) stated that the infestation may be seen anywhere in the world due to frequent overseas travel.

Spine is an unusual site with incidence of less than 1% of total hydatid disease burden (12). In contrast to visceral disease, bone or central nervous system involvement occurs much less frequently. Only 0.5 to 3.1% of patients suffer from bone involvement, half of which occurs in the spine (1, 5, 7, 23, 31, 35).

Spinal hydatitosis is a significant cause of spinal cord compression syndrome in endemic areas. Spinal disease most frequently involves the thoracic segments (46% to 50%) as in our cases all were of thoracic spine involvement. Lumbar (20% to 29%) and sacral (20% to 23%) (1, 4, 5, 14, 23, 28, 29, 30, 31, 34, 35, 39, 42). The cervical spine is the least commonly involved area.

Braithwaite and Lees classified spinal hydatid disease into five types (9).
1. Primary intramedullary hydatid disease
2. Intradural extramedullary hydatid cyst
3. Extradural intraspinal hydatid cyst
4. Hydatid disease of the vertebra
5. Paravertebral hydatid disease

The spinal involvement occurs in the form of cystic swelling with bone erosion causes back pain indistinguishable to Pott's spine, radiculopathy, myelopathy and pathological fracture. Immunological markers are not specific to the parasite and the specificity of ELISA for such antigen is about 50% due to cross reactivity by other parasitic antigens.

Spinal hydatid disease should be considered in the differential diagnosis of spinal cord compression syndrome in endemic countries. Due to the relative rarity of the entity most diagnoses are made intraoperatively which increases the risk of future recurrences. As the choice of the most suitable surgical approach relies on correct diagnosis and because special measures are required to prevent intraoperative spillage, preoperative diagnosis is of high importance.

Though plain skiagram and myelography were important tools few years back its utility in present era is limited. CT has excellent bone resolution and usually shows irregular erosions of the cancellous bone devoid of any subperiosteal reaction or enlargement of the vertebra (6, 23, 28, 29, 31).

The MRI with its superior soft tissue resolution is the most sensitive diagnostic method as well as the method of choice for determination of the extent of the disease (1, 8, 16, 29). A sausage-like shape with two dome shaped ends, thin and regular walls and no septation or debris in the lumen. The lesions are occasionally spherical. Signal characteristics of the cyst content are usually similar to that of CSF (2, 8, 29). On T1W images the cyst wall appears iso- or slightly more hypo-intense than the cyst content and enhances slightly after contrast injection. T2W images demonstrate a low intensity rim which correlates to the pathological findings of reactive fibrosis and degeneration surrounding the parasitic membrane. It has also been reported that viability of the cysts could be determined by their MRI characteristics (8). On T2W images cysts tend to lose their hyperintensity and the walls tend to lose their hypointense
signal upon inactivation (2, 8, 35). The few rare differentials are dorsal arachnoid diverticula, dorsal intrathoracic and lateral thoracic meningoceles.

Diagnostic fine needle aspiration is usually avoided because of the risk of spillage or anaphylaxis due to cyst rupture. Not uncommonly, the final diagnosis is delayed until the surgery and direct visualization of the lesions (6, 23, 39, 40).

Head, thorax and abdomen should be scanned for extra-spinal cysts upon the diagnosis of hydatid disease (16, 33). Surgery is currently considered the treatment of choice for spinal hydatid disease (1, 6, 8, 31). Thorough preoperative workup is essential.

The preferred surgical approach is posterior approach by laminectomy, decompression of neural tissue, removal of maximum possible cyst without spillage of their contents and debridement followed by three column fixation by means of pedicle screw and rod construct (11, 39).

The most common complication occurred during surgery is rupture of cyst. This is more common with large cystic hydatid rather alveolar hydatid cyst. The narrow path available in posterior approach makes this removal more complicated and time consuming. Author recommends patience during the whole procedure. The neurological manifestations progress more rapidly and are more severe with alveolar hydatid disease than with cystic hydatid disease. Alveolar hydatid disease is characterized by root pain and motor or sensory deficits below the level of the lesion. E. multilocularis cysts are small, group in clusters, elicit a severe inflammatory reaction from the host, and tend to metastasize both locally and distantly. The chances of inflammatory reaction can be reduced by padding the local tissue early during exposure. Bone destruction and pus-like fluid may be encountered during operation, leading to the name 'ossifluent abscess' (34).

Bone involvement is a major challenge for the treating surgeon. Due to the infiltrative nature of the lesion, spillage of scolices is unavoidable during the surgery for lesions involving the bone (6, 23, 28, 31). Even laminectomy may result in rupture of the cysts and spillage (23). It must be kept in mind that the disease is highly prone to recurrence, and plans for the surgical approach and stabilization must be made accordingly (43).

Simple decompression with laminectomy was by far the most commonly reported procedure (2, 14, 27, 44). This approach allows simple and easy decompression with little morbidity and mortality. However, access to the vertebral body is limited which may be a problem especially in recurrent disease. Stability of the anterior column must be ascertained to prevent postoperative kyphosis. Some authors advocate a more radical approach with as much bone removal as possible (23). Radical removal is not shown to preclude disease progression. However, extensive resection with stabilization and grafting is shown to prolong survival in selected cases (43).

Operations in thoracic cases were mainly posterior (4, 6, 7, 14, 18, 23, 31, 39). More extensive lesions with involvement of the vertebral body can be approached via a transpedicular route (7).

Drug therapy with benzimidazole derivatives is proven to be effective (13). A course of Albendazole (10-15mg/kg daily in divided doses) is continued for multiple cycles. Albendazole is preferred over Mebendazole due to better pharmacokinetic properties and superior efficacy against helminths (11, 22). While these agents do not cure the bone disease they are used in conjunction with surgery to decrease avoid recurrences due to intraoperative spillage (11, 22, 31, 39).

It should be kept in mind that both of these drugs are teratogenic and embryotoxic, and both may cause alterations in liver function and hematological adverse reactions (15). The complete blood picture and liver enzyme titre basal level as well, every monthly is assessed.

It is reported that the drug Praziquantel is also effective against Echinococci alone or in combination with Albendazole. Lam et al (22) reported an inoperable case of recurrent spinal hydatidosis treated with a combination of Albendazole plus Praziquantel having survived for 34 years.

CONCLUSION

Alveolar spinal hydatid disease is a rare condition. It is difficult not only to diagnose but to treat as well. The motto of this case series with various references is to educate readers about this rare entity that will help in future practice.

REFERENCES