Spinal Hydatid Cysts

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INTRODUCTION

Hydatid cyst can involve many systems, mainly the liver and lungs. The liver is the location of 50-70% of Echinococcus granulosus (EG) cysts while 10-30% are in the lungs and 10% in other regions of body. The spinal and cranial regions are rare areas of involvement. Bone involvement is seen in 0.5-4% (8). Bone lesions are most frequently localized at the spine and pelvis. There is no cyst membrane and unexplained fractures can be observed. It is an important public health issue since surgical treatment is challenging and the cyst does not respond well to medical therapy. Clinical findings are similar to those in other spinal pathologies. It may manifest with many diverse clinical findings ranging from simple low back pain to paraplegia. The most important principle is excision of the cyst without disrupting cyst integrity. Rupture during the first surgery will inevitably cause recurrences with multiple cysts (8, 10).

Epidemiology

Hydatid cyst is an endemic infectious disease in the Middle East, Australia, New Zealand, South America and China (0.3-0.5%) (10). It is due to excessive consumption of sheep and cattle, which are intermediate hosts of hydatid cyst. In addition, consumption of raw cattle products also has a major contribution to the endemic disease. Cyst hydatid is generally observed in children and young adults. There is no gender preponderance (1). It is most frequently seen at the fourth decade.

Pathogenesis

Hydatid cyst is a zoonotic disease caused by Echinococcus granulosus. The larval stage of E. granulosus is a metacestode that is seen as hydatid cyst. The definitive host is generally dogs. Intermediate hosts include sheep, cattle, swine, goat and monkey. Humans are also considered an intermediate host. Transmission most frequently occurs via the fecal-oral route. The organism can pass to the intermediate host by ingestion of water and food contaminated by the feces of the definite host. Subsequently, it attaches to the intestinal mucosa and reaches the liver and lungs through the portal vein circulation. Larvae spread to the whole body through the superior vena cava. The cyst contains clear fluid with a density of 1007-1015 (22).

Classification

Spinal hydatid cysts were classified into 5 groups by Braitwade and Less in 1981 (5). This classification has been used for over thirty years. In this classification, hydatid diseases are classified as follows:
Spinal Hydatid Cysts

- Primary intramedullary
- Intradural extramedullary
- Extradural intraspinal
- Intravertebral
- Paraspinal

The dumbbell formation type was added to the classification in recent years. Intraspinal and paraspinal disease are the most commonly seen forms (17).

**Frequency and spread of spinal hydatid cysts**

Cystic lesions are most commonly observed in the liver (70%) and lungs (20%). They are also seen in bones at a rate of approximately 0.5-4% (18). Kammere et al. reported that hydatid cysts of the central nervous system comprise 3% of all hydatid cyst cases and more than 50% of these cases are localized at the spinal system (15). Primary bone involvement occurs at a rate of 0.5-2% in non-endemic regions while this rate can be doubled in endemic regions (13).

In the spinal region, hydatid cysts are most commonly seen at the thoracic region (45-50%), followed by the lumbar region (20-39%). It is thought that the most common reason for this involvement pattern is vascular spread in spinal metastasis (1,7). Sacral and cervical involvement is extremely rare.

Larvae of echinococcus become localized at the trabecular tissue initially and then spread to cortical regions of the bone. Pathological compression fractures can be seen in the vertebrae with weakened trabecular and cortical areas. The discs serve as a natural barrier against spread of infection as in other primary spinal infections. However, this feature disappears in case of advanced infection.

Extradural localization in nine of 10 patients with general involvement of the corpus vertebra is reported in the literature (13). Primary intradural involvement is extremely rare. Intadural hydatid cyst generally occurs as a complication of surgery.

**Clinical presentation**

The most common complaints are back and low back pain. A pain pattern related to spinal deformity can develop in advanced cases. Neurological findings can vary depending on the localization of the cysts. Although neurological examination is normal in most cases, cauda equina or paraplegia can be seen in some cases (6, 7, 13).

Some studies reported that lower extremity weakness with low back pain is the most frequent finding (4, 14).

Although spinal hydatid cyst is a parasitic infection, its behavior resembles a malignant tumor. Given the clinical symptoms, spread pattern and limited response to treatment, the cases where total excision has failed in the first surgery tend to recur as in benign tumors and cause distant metastases as in malignant tumors.

**Diagnosis**

Since the disease presents with many diverse symptoms and findings that can produce either benign or severe disease presentations, and may be chronic, subacute or acute that requires immediate intervention, it causes low level of suspicion in non-endemic regions. The history, physical examination, radiological assessment, and serological tests are helpful in the diagnosis. Computerized tomography (CT) and magnetic resonance imaging (MRI) are as valuable as serological evaluations in both the diagnosis and follow-up. It is impossible to distinguish Taenia and Echinococcus eggs in the stool of definitive or intermediate hosts by light microscope. ELISA is currently one of the most frequently used methods and detects cystic coproantigens. The serological response against echinococcal antigens can vary depending on the host and localization of the cyst. Echinococcal antigens and antibodies can be used in the diagnosis. Several techniques including indirect hemagglutination (IHA), indirect immunofluorescence (IFAT), latex agglutination, solid-phase radioimmunoassay, immunoelcrophoresis, counter electrophoresis, Western Blot and ELISA can be used to detect anti-echinococcal antibodies while co-agglutination, counter-current immunoelcrophoresis and ELISA are used to detect echinococcal antigens. Antigens persist for several years after surgical resection and it is more appropriate to study echinococcal antigens in order to determine active or new infection. IHA can provide false-positive results due to shared antigens with Taenia solium, Taenia saginata, Ascaris lumbricoides, Fasciola hepatica and Plasmodium spp. Thus, titers ≥1: 360 are considered to be significant in the diagnosis of hydatid cyst (18). In addition, polymerase chain reaction (PCR) is also used in the diagnosis (16). The sensitivity and specificity of serological tests vary depending on organ involvement. Identification of DNA isolated from the stool or eggs by PCR is an alternative method.

Eosinophilia (20-35%) can be present on complete blood count due to the allergic features of the cyst. Sensitivity of
serological techniques is 80-90% for hepatic hydatid cyst but can be as low as 20-50% in spinal involvement (16). It has been reported that the sensitivity and specificity of the tests used in serological diagnosis depend on the features of the antigen used, the host from which the antigen is derived, the antibody response of the patients, the method used and the anatomic localization (11).

**Imaging**

On plain radiography, *Echinococcus granulosus* related changes in the spinal column do not differ from those seen in brucellosis, tuberculosis and other spinal infections. MR imaging is the gold standard for the radiological diagnosis of hydatid cyst cases. Heterogeneous hypo-intense cysts are seen on T1-weighted images while heterogeneous hyper-intense areas are observed on T2-weighted images (Figure 1, 2). Since this is a chronic disorder, findings due to bone erosion and expansion can be observed as in other benign pathologies. Bone erosion and related findings can be seen on CT scan (9).

**Differential diagnosis**

Arachnoid cyst, Pott’s disease, brucellosis, syphilis, cysticercosis, osteomyelitis, multiple myeloma and hyperparathyroidism should be considered in the differential diagnosis of spinal hydatid cyst (19, 20).

**Management**

Hydatid cyst is a zoonotic disease that has been reported since Hippocrates and is difficult to treat. Antihelminthical agents are used in the treatment (2). However, it is emphasized that drug therapy is inadequate for cure and should be supported by surgical treatment (21). Medical therapy is initiated as soon as possible after diagnosis. Albendazole, mebendazole and praziquantel are antihelminthical agents used in the treatment of hydatid cyst. Albendazole is preferred in the treatment of spinal hydatid cyst due to its absorption and efficiency. It is used at a dose of 800 mg/day (twice daily). Hematological disorders and changes in liver enzymes can be observed with albendazole and mebendazole, as with many other drugs (12). The effectiveness of medical therapy is monitored by serology and imaging studies (3). Praziquantel alone has a low success rate in the treatment; thus, it is recommended to use praziquantel in combination with albendazole in the preoperative period. When the praziquantel plus albendazole combination is used, doses are 50 mg/kg/day and 10-15 mg/kg/day for a month, respectively (1-4).

**Surgical treatment**

Since EG cysts contain scolexes, the basic principle in surgery is to remove the cyst without disrupting its integrity. Irrigation with 10% povidine or saline solutions should be performed for at least 15-20 minutes in cases with cyst rupture or against the risk of cyst rupture. En bloc resection of the cyst provides cure in primary spinal hydatid cyst. Otherwise, cysts spreading to the epidural area will cause recurrence that is impossible to treat by surgery. When
Spinal Hydatid Cysts

CONCLUSION

Hydatid cyst is a serious parasitic infection worldwide, especially in developing countries. Many agents used in parasitic infections fail to achieve adequate treatment success. Although spinal hydatid cyst is a rare disease, it is a serious problem due to lack of sufficient response to medical therapy and challenges in surgical excision. The fact that cure is impossible by surgery in cases with recurrence highlights the importance of radical planning in the first surgery.

REFERENCES


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