

# Spinal Hydatid Cyst: Review

## Spinal Hidatik Kist

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**ABSTRACT** Hydatid cyst infection is one of the oldest diseases known in the history of humanity. It is a disease caused by the larval *Echinococcus granulosus* tapeworm and is seen in regions where parasitic infestations are endemic. Spinal hydatid disease is an infestation characterized by multivesicular diffuse infiltration of the cancellous bone, including corpus vertebrae, pedicles, and the lamina. This is a rare condition leading to severe neurological problems. In the spinal area, the most commonly involved part is the thoracic region. Diagnosis is difficult and frequently delayed until signs and symptoms of spinal cord and nerve compression develop. There are no characteristic signs and symptoms. Most common signs are low back pain, radicular pain, paraparesis or paraplegia, sensation disorders, and bladder/intestine dysfunction. Magnetic resonance imaging (MRI) is the most sensitive diagnostic method. In the treatment of spinal hydatid cyst, surgery is still accepted as the "gold standard". Anthelmintic agents and benzimidazole derivatives can also be combined with surgical treatment, particularly for preventing recurrences. After review of the related literature, it is emphasized that physicians should be careful in the differential diagnosis of patients applying with spinal pain, radicular pain, progressive myelopathy or spinal cord compression syndrome. Because of high mortality and morbidity; importance of early diagnosis and treatment is highlighted along with emphasizing eradication of disease.

**Key Words:** Echinococcosis; spinal cord compression; spinal diseases

**ÖZET** Hidatik kist enfeksiyonu, insanlık tarihinde bilinen en eski hastalıklardan biridir. *Echinococcus granulosus* adlı tenyanın larvalarının neden olduğu bir hastalıktır ve parazitik enfestasyonların endemik olduğu bölgelerde görülür. Spinal hidatik hastalık korpus vertebra, pediküller ve laminaları kapsayan, süngerimsi kemiğin çoklu keseciklerle yoğun infiltrasyonu ile kendini gösteren bir enfestasyondur. Bu, ciddi nörolojik problemlere yol açabilen, nadir görülen bir durumdur. Spinal alanda en sık tutulan bölüm toraks bölgesidir. Tanısı zordur ve sıklıkla spinal kord basısı ve sinir basısına ait bulgu ve semptomları ortaya çıkıncaya kadar tanı konması gecikmektedir. Buna özgü bir bulgu ve semptom yoktur. En sık bulguları bel ağrısı, radiküler ağrı, paraparezi veya parapleji, duyu bozuklukları ve mesane/bağırsak bozukluğudur. Manyetik Rezonans Görüntüleme (MRG) en duyarlı tanı metodudur. Spinal hidatik kistin tedavisinde, cerrahi halen "altın standart" olarak kabul edilmektedir. Antihelmintik ajanlar ve Benzimidazol türevleri de özellikle, rekürensleri önlemek için cerrahi tedavi ile birleştirilebilir. İlgili literatürün gözden geçirilmesiyle, spinal ağrı, radiküler ağrı, progresif miyelopati veya spinal kord kompresyon sendromu ile başvuran hastaların ayırıcı tanısında doktorların dikkatli olması gerektiği vurgulanmaktadır. Yüksek mortalite ve morbidite nedeniyle, hastalığın eradikasyonu üzerinde durulurken, erken tanı ve tedavinin önemine dikkat çekilmektedir.

**Anahtar Kelimeler:** Ekinokokkozis; spinal kord kompresyonu; omurga hastalıkları

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Hydatid cyst infection is one of the oldest diseases known in the history of humanity.<sup>1</sup> Hydatid cyst is a disease caused by the larval *Echinococcus granulosus* tapeworm<sup>2-5</sup> and is seen in regions where parasitic infestations are endemic.<sup>4,6</sup> Animals are main culprits in the spread

of hydatids and the disease is seen in all the countries of the world, and especially in the Mediterranean countries, Turkey, Africa, Middle Eastern and Asian countries, East and South Europe, Latin and South America, Australia, and New Zealand.<sup>1,4,7</sup> The definitive hosts in nature are dogs, wolves, and other carnivores, becoming infected by eating the raw giblets containing the cyst. The adult parasite lives in the intestine of the host and its eggs pass into the feces to spread to the environment. Many animals such as the sheep, goat, cattle, camel, horse, and pig are intermediate hosts, and when they take the egg, embryos are formed in the intestine to enter circulation via the venous and lymphatic route. Humans can be intermediate hosts, they can be infected by direct contact with the definitive host, or by orally taking water, food, and green vegetables contaminated with the feces of the definitive host.<sup>5,7</sup> The embryos enter circulation following their penetration into the intestine, spread to the portal circulation leading to cyst formation in visceral organs or destruction by the host.<sup>5</sup>

Spinal hydatid disease was first defined by Churrier in 1807<sup>5,6,8</sup> and the first surgical operation was reported by Reydellet in 1819.<sup>5,8</sup> *E. granulosus* mostly involves the liver (50-60%) or lungs (20-30%), however any body organ (i.e., brain, heart, bones) can also be involved (< 10%).<sup>4,7</sup> Only 0.5-3.1% of patients have bone involvement and half of them have concomitant spinal involvement.<sup>8-11</sup>

Spinal hydatid disease is an infestation characterized by multivesicular diffuse infiltration of the cancellous bone, including corpus vertebrae, pedicles, and the lamina, and the disease is caused by the larvae of *E. granulosus*.<sup>8</sup> Its incidence is 1.1% in Australia and 14% in Tunisia. In endemic countries it is an important cause of spinal cord compression syndrome.<sup>4,8,9,12-15</sup> In Turkey, it was reported that spinal hydatid disease was responsible for 3.8% of all cases with spinal cord syndrome<sup>4,8,9,12-15</sup> and in 5% of cases operated with the diagnosis of spinal cord compression, hydatid disease was present.<sup>16</sup>

In the spinal area, the most commonly involved parts are the thoracic region (46-50%) while the lumbar (20-29%) and sacral (20-23%) segments

are involved less frequently. Involvement of the cervical region is reported to be the least common.<sup>4,8,9,12,14,15,17</sup>

Spinal involvement was classified by Braithwaite and Lees into 5 types: 1. Primary intramedullary, 2. Intradural extramedullary, 3. Extradural intraspinal, 4. Vertebral, and 5. Paravertebral involvement.<sup>18</sup> The first 3 types are rare.<sup>5</sup>

It is thought that the infestation primarily starts at the center of the vertebral body and spreads through extradural or paravertebral route. Cases with involvement of the bone and epidural space constitute 90% of patients. There is intradural extramedullary involvement in 9% of cases.<sup>4-6,8,9,15,19,20</sup> In the literature, intraspinal extradural and/or paraspinal localization of hydatid cyst cases are very frequently reported,<sup>6,13,15,18,20-25</sup> intradural cases have also been reported.<sup>1,3,26-28</sup> Compression and cortical weakening secondary to osteonecrosis lead to pathological fractures and spreading to the pedicle, lamina, and costae can take place.<sup>4,5,17,20</sup> As the cyst development was limited with the periosteum, intervertebral disc spaces are generally preserved.<sup>4,5,20</sup>

The rate of cyst development is 7 mm/month.<sup>9</sup> Because of the slow progression of the cyst and the resistance of the surrounding bone, it takes a long time for the clinical manifestations to crop out. There are no characteristic signs and symptoms. The rate of some symptoms are as follows: back pain 85%, radicular pain 25-95% and paraparesis 25-77%.<sup>4,17</sup> Another study stressed out that the most common signs were paraparesis (62%) or paraplegia (26%), low back pain or radicular pain (55%), sensation disorders (36%), and sphincter deficits (30%).<sup>5</sup> In a meta-analysis performed in our country, the rates of common signs were as follows: weakness in lower extremities 73%, low back pain 43%, bladder/intestine dysfunction 32%, pain in lower extremities 27%, sensation disorders 24%, and paravertebral swelling 2%.<sup>20</sup>

Most cases reported with spinal hydatidosis are males (62%) and their ages range between 6-74 years (mean 36 years). Because of its slow progression, the disease is primarily seen in adults.<sup>5</sup> Cases in childhood have been reported rarely.<sup>10,17,23,26,29,30</sup>

Diagnosis is difficult and frequently delayed until signs and symptoms of spinal cord and nerve compression develop. The best approach is a detailed history and clinical examination combined with imaging techniques.<sup>5,17</sup> Laboratory tests other than eosinophilia are not helpful. In abdominal disease, sensitivity and specificity of ELISA, indirect hemagglutination, and complement fixation tests are 80-100% and 88-96%, respectively. However, in extrahepatic involvement their sensitivity decrease (25-56%); their value in the diagnosis and follow-up of primary bone involvement is limited. In spinal hydatid disease, imaging techniques are more sensitive than serodiagnosis. In the presence of confirmative imaging techniques, a negative serology does not exclude the diagnosis.<sup>7</sup> Due to risk of spread by cyst rupture and anaphylaxis, diagnostic needle aspiration biopsy should be avoided. Diagnosis can be delayed until direct observation of lesions during operation.<sup>4,8</sup>

In direct roentgenograms, although not diagnostic, “moth eaten” lesions with the surrounding sclerosis and calcifications extending to the paravertebral soft tissue confirm the diagnosis.<sup>4,15,31</sup> Changes in bone structure are reported in only 27% of cases. Shortening in the height of vertebra corpus and compression fractures or pedicle erosion can be striking.<sup>4</sup> Myelogram has little diagnostic value and should be avoided because of intradural spread secondary to cyst rupture or anaphylaxis.<sup>4</sup> In computerized tomography (CT), irregular bone erosions are usually seen without any subperiosteal reaction and a vertebral expansion.<sup>4,8,12,14</sup> Magnetic resonance imaging (MRI) is the most sensitive diagnostic method.<sup>4,9,21</sup> CT and MRI are beneficial both in diagnosis and follow-up. Imaging techniques are important in terms of early and accurate diagnosis.<sup>3,4,32</sup> Fahl et al. defined MRI and stated that cysts usually look like flattened sausages and have two dome-shaped ends, consisting of thin and regular walls and not including septa or debris within the lumen.<sup>33</sup> Lesions sometimes are spherical. Intradural cysts can be single or multiple, however, extradural cysts are always multiple and involve the bone. Signal characteristic of the cyst content is usually similar to that of cerebrospinal

fluid.<sup>21</sup> MRI of a patient with paraplegia associated with spinal hydatid cyst followed up in our rehabilitation clinic is showed in Figures 1 and 2.

In the differential diagnosis, vertebra tuberculosis, pyogenic infections, fibrous dysplasia, enchondroma, malignancies, multiple myeloma, giant cell tumors, and hyperparathyroidism should be kept in mind.<sup>4,5,34</sup> In tuberculous spondylitis, involvement in one or more vertebral segment(s), intervertebral disc destruction, enlarged and calcified paravertebral mass, along with the absence of sclerosis is noticeable. L1 is the most commonly affected vertebra. Infection generally starts at the anterior part of the vertebral corpus, related with the subchondral bone. Tuberculosis can rarely lead to an extradural granulomatous lesion without associated bone involvement. This is mostly seen in the thoracic region and in the epidural space.<sup>34</sup> Normelli et al. reported a 36 years old Iraqi male, who was operated three times for progressive paraparesis and thoracolumbar hydatid cyst, and this patient developed destruction at T10-L1 level with a 45° gibbus deformity.<sup>31</sup> The resemblance of this case with tuberculous spondylitis is striking. Misdiagnosis can frequently be the case.<sup>5,17,20</sup> The initial diagnoses can be Pott’s disease<sup>13,17,20</sup> spinal tumor<sup>5,20</sup> or disc herniation.<sup>20</sup> A study from Turkey revealed that 17% of patients were misdiagnosed as Pott’s



**FIGURE 1:** T2W axial images of our patient with hydatid cyst showing a lobulated contoured cystic lesion extending to the spinal canal and compressing the spinal cord.



**FIGURE 2:** T2W sagittal images of our patient showing extradural hydatid cyst which was sausage-like shape in thoracic spine.

disease.<sup>20</sup> Brushi et al. reported a Tunisian patient with progressive lumbosciatalgia in whom a misdiagnosis of Pott's disease led to administration of triple drug therapy as antituberculous treatment.<sup>13</sup> In Turkey, in the last five decades, 28 studies presented as spinal hydatid cyst were investigated and a total of 84 cases with intraspinal extradural localization and thoracic and vertebral involvement were determined. Initially, 14 cases were misdiagnosed as Pott's disease, 13 as spinal tumor, three as disc herniation, and seven were accurately diagnosed as hydatid cyst, while in 47 cases no prediagnosis could be made.<sup>20</sup> Schnepfer et al. in North America presented a 34 years old Turkish woman with recurrence of thoracic extradural disease, which was misdiagnosed and ineffectively treated.<sup>5</sup> She had paraparesis, and together with the MRI findings, the prediagnosis favored spinal cord tumor, however, after four years, the symptoms re-

curred, the patient was re-operated for the cysts and albendazole treatment was started.

In the treatment of spinal hydatid cyst, surgery is still accepted as the "gold standard".<sup>4,5,7,8,35</sup> As most cases apply with signs and symptoms of spinal cord compression, surgery is performed. In general, posterior approach is preferred. Laminectomy along with simple decompression is the most common procedure. When there is bone involvement, possibility of rupture during surgery is high due to the infiltrative nature of the lesion. The high tendency of recurrence should always be kept in mind and the surgical approach and stabilization should be planned accordingly.<sup>4,5</sup> With the aim of preventing the spread secondary to cyst rupture, before the resection, a cysticidal agent (hypertonic 30% saline, cetrimide, or 70-95% ethanol) can be given together with some amount of fluid in order to destroy the cysts, however this method has not been validated by clinic trials.<sup>4,7</sup>

Anthelmintic agents and benzimidazole derivatives can also be combined with surgical treatment, particularly for preventing recurrences. Especially albendazole is preferred over mebendazole as it has better gastrointestinal absorption, with higher plasma levels.<sup>7,36</sup> Following surgery, 800 mg/day in two divided doses and for three or more cycles (four weeks of drug treatment, two weeks of drug-free period) are recommended for a total of 3-12 months.<sup>7,16</sup> These drugs may have teratogenic and embryotoxic side effects, as well as adverse effects on liver and hematological systems.<sup>7</sup>

Recurrences lead to multiple operations.<sup>1,17,37,38</sup> Because of high recurrence rates, it has a poor prognosis and has been compared to spinal malignancy.<sup>1,4,5,17,20,37</sup> The rates reported in the literature range between 30% and 100%.<sup>4,5,10,17,37</sup> When there is recurrence, the rate of paraplegia increase to 45%.<sup>5</sup> Turtas et al. reported the recurrence rate as 100% in the 11 cases they followed-up for 20 years, and the mean number of surgical procedures per patient was reported as 4.8.<sup>38</sup> They emphasized that recurrence increased when treatment was delayed. After the operation, recurrence can be seen in 2-28 months (mean 25.2 months). Rate of recurrence was lower in intradural extramedullary



involvement.<sup>4,8</sup> Mortality rates vary and range between 3% and 50%.<sup>10,17,37</sup> Operative mortality rate is 15% and increases with repetitive surgeries.<sup>4</sup> With early and accurate diagnosis, early surgery improves the prognosis.<sup>2,27</sup>

## CONCLUSION

Spinal hydatid cyst can be seen in non-endemic countries because of people traveling all over the world. Despite modern surgery and pharmacological treatment, it still is hard to recover this disease

with a high tendency for recurrence. Mortality and morbidity is high because it leads to progressive destruction in the vertebral column and neurological problems. Physicians should be careful in the differential diagnosis of patients applying with spinal pain, radicular pain, progressive myelopathy, or spinal cord compression syndrome. Although diagnosis is difficult, the best approach is a detailed history and clinical examination combined with imaging techniques. The best treatment is preventing the disease.

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