

## HYDATID CYST OF THE CRANIOCERVICAL JUNCTION: CASE REPORT

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**OBJECTIVE AND IMPORTANCE:** Although rare in developed countries, hydatid disease is a serious parasitic infection in endemic areas. Spinal disease most commonly involves the thoracic region, and involvement of the upper cervical spine is extremely rare. In this report, a case of hydatid disease involving the craniocervical junction is presented, along with a review of the literature.

**CLINICAL PRESENTATION:** A 44-year-old man presented with the complaint of neck pain. The results of his physical and neurological examinations were within normal ranges, except for pain exacerbated by neck motion and spasm of the cervical musculature. Magnetic resonance imaging demonstrated a cystic lesion involving the odontoid process and body of C1 and C2, with thin and regular cyst walls and cyst contents similar in intensity to that of cerebrospinal fluid. The results of serological tests performed with the suspected diagnosis of hydatid disease were positive.

**INTERVENTION:** The patient initially underwent surgery to provide stabilization of the craniocervical junction, using autogenous bone graft and sublaminar wiring from the occiput to C3 via a posterior approach. The cyst was approached via a transoral route, using a U-shaped pharyngeal incision. There were no neurological deficits after surgery. Postoperative magnetic resonance imaging scans confirmed complete excision of the cyst, and the patient was discharged on the 12th postoperative day. He received six cycles of albendazole treatment, each consisting of 28 days with an intervening drug-free period of 2 weeks. Magnetic resonance imaging scans performed 1 year after surgery revealed the patient was still disease-free.

**CONCLUSION:** Hydatid disease should be considered in the differential diagnosis of spinal cord compression, especially in endemic areas. Although the chance of obtaining a cure is unlikely, radical surgery coupled with antihelminthic therapy seems to provide long-lasting relief.

**KEY WORDS:** Craniocervical junction, *Echinococcus*, Hydatid cyst, Spine

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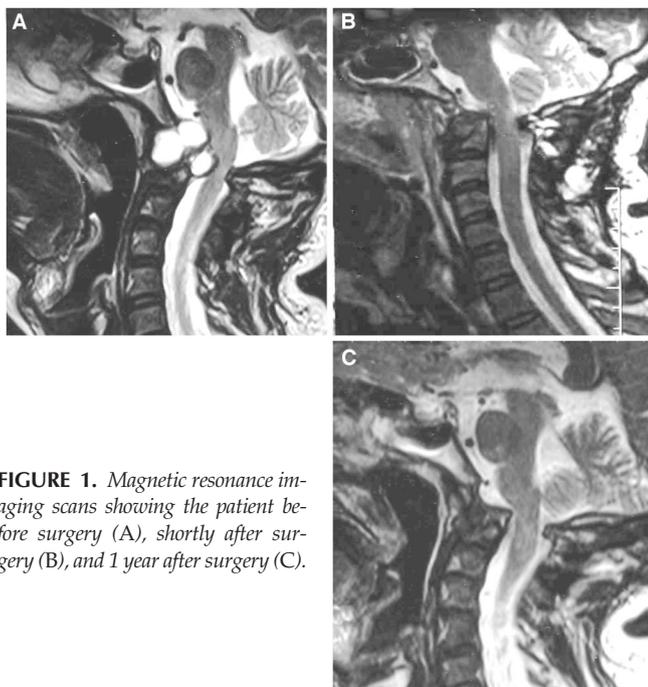
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Although rare in developed countries, hydatid disease is a serious parasitic infection in endemic areas such as Turkey (1). Bone involvement occurs in 0.5 to 2% of cases, with 44% of these cases involving the spine (13). Five percent of the operations performed with the diagnosis of spinal cord compression in Turkey are attributable to hydatid disease (8). Spinal disease exclusively involves the thoracic region. Involvement of the upper cervical spine is extremely rare (7). There is only one case reported in the literature involving the craniocervical junction (3). In the current report, a case of hydatid disease involving the craniocervical junction is presented, along with a review of the literature.

### CASE REPORT

A 44-year-old man presented with the complaint of neck pain. The results of his physical and neurological examina-

tions were within normal ranges, except for pain exacerbated by neck motion and spasm of the cervical musculature. A routine cervical x-ray was performed for the initial evaluation. The contours of the odontoid process could not be appreciated; therefore, evaluation proceeded with computed tomography of the craniovertebral region, which revealed a cystic lesion eroding and expanding the odontoid process and the involvement of the C1 and C2 vertebral bodies. Magnetic resonance imaging demonstrated a multilobular cystic lesion with thin and regular cyst walls. The cyst contents were hypointense on T1-weighted images and hyperintense on T2-weighted images, with an intensity similar to that of cerebrospinal fluid, and there were no septations within the cyst (Fig. 1A). Serological tests performed with the suspected diagnosis of hydatid disease confirmed the diagnosis. Albendazole (400



**FIGURE 1.** Magnetic resonance imaging scans showing the patient before surgery (A), shortly after surgery (B), and 1 year after surgery (C).

mg administered twice daily) was initiated accordingly. Routine systemic evaluation did not reveal the involvement of any other organs, including the liver.

The initial concern was stability before excision of the lesion. Therefore, a stabilization process using autogenous bone graft and sublaminar wiring from the occiput to C3 was performed via a posterior approach. Hyperflexion, hyperextension, and rotational films confirmed the stability of the craniocervical junction after surgery.

The cyst was approached via a transoral route, using a U-shaped pharyngeal incision. The cyst was ruptured during excision, and copious irrigation of the surgical field was performed using hypertonic saline solution. A tracheostomy was performed before completion of the procedure.

The patient was followed up in the intensive care unit for 5 days. He received broad-spectrum antibiotics in addition to albendazole and was fed with a nasogastric tube. There were no neurological deficits after surgery. The tracheostomy was closed on the fifth postoperative day, and the patient was transferred to the ward. Histopathological examination of the cyst wall confirmed the diagnosis of a hydatid cyst, revealing an outer laminated nonnucleated layer and an inner germinative layer. Postoperative magnetic resonance imaging scans confirmed the complete excision of the cyst (Fig. 1B). The patient was discharged on the 12th postoperative day.

The patient received six cycles of albendazole treatment, each consisting of 28 days with an intervening drug-free period of 2 weeks. Magnetic resonance imaging scans performed 1 year after the surgery revealed the patient still to be disease-free (Fig. 1C).

## DISCUSSION

Hydatid disease is a significant parasitic infection in endemic areas such as Turkey; however, contrary to common belief, the incidence has not decreased significantly during the past decade (1). Spinal hydatid disease was first described by Churrier in 1807, and the first surgical operation was performed by Reydellet in 1819 (11). Today, 5% of the operations performed with the diagnosis of spinal cord compression in Turkey are attributable to hydatid disease (14).

Hydatid disease is caused by the larval stage of the tapeworm, *Echinococcus granulosus*. The definite host is generally the domestic dog. The gravid proglottid excreted in the feces of the dog ruptures to release ova, which are accidentally ingested by the intermediate hosts, one of which is humans. After ingestion, oncospheres develop in the duodenum and penetrate the mucosa to enter the portal circulation and disseminate throughout the body (8).

The most commonly involved site is the liver, and then the lung; bone involvement occurs in 0.5 to 2% of cases, 44% of which involve the spine (13). Infestation begins in the center of the vertebral body and extends into the epidural space; therefore, the mass is epidural in 90% of cases (7). The thoracic region is the most commonly involved spinal site, and then the lumbar region (14). The upper cervical spine is rarely involved. There is only one case reported in the literature involving the craniocervical junction (3). In that report, multicystic hydatid disease involving the upper cervical spine as well as the craniocervical junction was present. The surgical pathological finding was extension of one of the cysts paravertebrally into the cervical musculature. The patient was operated on by an otorhinolaryngologist, and there was no neurosurgical intervention. The currently presented case is the first case of hydatid disease involving the craniocervical junction in the neurosurgical literature.

The treatment strategy of hydatid disease depends on aggressive surgery combined with antihelminthic therapy (10). Reported procedures involving the cervical region generally involve posterior approaches with a laminectomy (15). Keller et al. (6) described the case of a cervical hydatid cyst treated with an anterior corpectomy, bony fusion, and external halo bracing. The patient was reported to develop progressive kyphosis and myelopathy. Our experience has led us to think that the best decompression can be obtained anteriorly and the best stabilization can be achieved posteriorly. Therefore, the patient was first approached posteriorly, and stabilization was performed from the occiput to C3, using sublaminar wiring and autogenous costal bone graft. The cyst was excised via the transoral route. Although their effectiveness is not proven, many surgeons prefer to use scolecoidal agents, such as 3% hypertonic saline, after removal of the cyst, with the hope of preventing recurrence (12). Hypertonic saline was used for irrigation of the surgical field in the present case.

Albendazole is the preferred antihelminthic in the treatment of hydatid disease (8). Its use is advocated, in cycles, for 3 months to 1 year after surgery, and indefinitely if recurrence

occurs (14). There are recent articles reporting the effectiveness of solitary albendazole treatment for surgically inaccessible lesions (2). It is also reported to be effective in prolonging patient survival in inoperable cases (9). However, surgery is still the “gold standard” in the treatment of spinal hydatid disease (10).

Obtaining a cure is unlikely, however, and reoperations are generally needed (4). In a series of 11 patients followed for 20 years, the recurrence rate was 100% and the average number of surgical procedures for a single patient was 4.8 (15). In a more recent article, the average length of remission after radical surgery coupled with antihelminthic therapy was 29.6 months (10). The mortality rate is 0 to 3% for the initial operation and progressively increases to 15% in reoperations (11). Conversely, Garcia-Vicuna et al. (5) stated that they have initiated albendazole therapy in a patient with cervical spinal hydatid disease after surgery and never ceased therapy but continued in a cyclical fashion. The patient was reported to be disease-free after 9 years.

Hydatid disease should be considered in the differential diagnosis of spinal cord compression, especially in endemic areas. Although the chance of obtaining a cure is unlikely, radical surgery coupled with antihelminthic therapy seems to provide long-lasting relief.

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## COMMENTS

The authors present a case of hydatid cyst with compression of the cervicomedullary junction. This is a serious parasitic infection in anemic areas and approximately 5% of spinal cord compression is caused by hydatid cyst disease in Turkey. The patient was appropriately treated by initial occipitocervical stabilization and subsequent transoral resection and medical therapy.

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The authors present an unusual case of hydatid disease involving the cervical spine. Although this disease is uncommon in the United States, it is endemic in many parts of the world. Spinal hydatid disease usually occurs in the thoracic region and presents with spinal cord compression. The case presented was aggressively treated with a posterior fusion followed by a transoral resection of the cyst. One of the strengths of this article is that the authors emphasize the necessity for surgical resection of these cystic lesions. As with other aggressive infectious processes that involve the central nervous system, such as fungal infections, neurosurgical intervention plays a major therapeutic role in the care of these patients. Even though antihelminthic treatment has improved with new agents such as albendazole, medical management alone is insufficient therapy for spinal hydatid disease and rapid surgical decompression is imperative to prevent potential catastrophic neurologic morbidity.

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The authors bring to our attention a disease with which most American neurosurgeons have limited, if any, experience. Their surgical management may serve as a reference to those who might be confronted with a similar patient. The addition of antihelminthic drug treatment (albendazole) seems to alter the previously reported 100% recurrence rate in patients with spinal involvement (3). The authors cite two articles, in addition to their own article, suggesting that antihelminthic drug therapy is beneficial in preventing recurrences in cases in which total cyst removal cannot be accomplished (1, 2).

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