Diagnosis and treatment of multiple hydatid cysts at the craniovertebral junction

Case report

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✓Echinococcosis, or hydatid disease, of the craniovertebral junction and skull base is rare. The authors report the occurrence of multiple hydatid cysts at this anatomical location in a young woman who was previously misdiagnosed with tuberculosis. The patient underwent transoral excision of the hydatid cysts followed by posterior decompression and occipitocervical fusion. She was treated postoperatively with albendazole for 12 weeks with a good outcome. The management of spinal hydatid disease is reviewed.

KEY WORDS • echinococcosis • craniovertebral junction • hydatid cyst

ECHINOCOCOSIS, also known as hydatid disease, is caused by infection from the larval form of the Echinococcus genus.12 Humans are the intermediate hosts and get infected through the fecal–oral route. The common infection sites involved are the liver, lungs, and brain. Primary hydatid disease involving the spine is rare, even in regions endemic to this disease, and constitutes only 1% of the disease spectrum.2,3,7,9,14,17–19,21–23 Moreover, spinal hydatid cysts can be confused with tuberculous cold abscesses of the spine.3,7,25 Spinal hydatid disease has been reported in the cervical,10,16 thoracic,2,3,17,22 lumbar,15,17 and sacral regions.21 There have been three case reports of multiple hydatid cysts involving the CVJ.4,7,23 We report a young woman with multiple primary hydatid cysts at the CVJ who presented with severe neck pain and neurological deficits. The cysts were excised using anterior transoral and posterior approaches followed by occipitocervical fusion. She was given albendazole postoperatively for 12 weeks.

Case Report

History and Examination. This 23-year-old woman had been experiencing neck pain, restriction of neck movement, paresthesia in the upper limbs, and mild weakness of the upper limbs for 3 years prior to presentation at our hospital. She also had severe neck pain when moving her neck. On the basis of imaging results, tuberculosis of the CVJ was diagnosed and she received antituberculous chemotherapy for 16 months with no improvement in her symptoms. At presentation to our hospital she had clinical features of an upper cervical cord compression. The results of her cranial nerve examination were normal. There was a mass seen in the posterior pharyngeal wall of the patient. Magnetic resonance imaging of the cervical spine showed multiple cystic lesions encircling the entire CVJ, involving the C-1 anterior arch and C-2 body (Fig. 1A). The lesions were hypointense on T_1-weighted images and hyperintense on T_2-weighted images (Fig. 1B and C). The cysts were causing compression of the cervicomedullary junction.

Operation. The patient underwent a transoral procedure during which the posterior pharyngeal swelling was initially aspirated and clear fluid was found. This clear fluid
strongly indicated a possibility of hydatid disease; therefore 3% hypertonic saline was used to irrigate the operative site and was injected into the cysts. After the mucosa were incised, a classic milky white cyst wall was encountered, confirming our suspicion of a hydatid cyst. The cysts were excised during concurrent steroid therapy to avoid an anaphylactic reaction. The anterior arch of C-1 and the remaining part of the dens were removed, and the cysts posterior and superior to this area were excised. The entire operative field and surrounding regions were irrigated with hypertonic saline. The mucosal wound was closed in layers.

Subsequently the patient was positioned prone and the posterior arch of C-1, part of the C-2 lamina, and the rim of the foramen magnum were exposed. There were cysts with involvement of the right half of the C-2 lamina and posterior arch of C-1, and these were both removed. There were cysts posterolaterally on the right side that were also excised. An occipitocervical fusion using a titanium Ransford loop and Atlas cables (Medtronic Sofamor Danek) was performed from the occiput to C-3 (Fig. 2). An iliac bone graft was used for bone fusion.

Postoperative Course. The biopsy specimen results confirmed the diagnosis of hydatid cysts (Fig. 3). Chest radiography and ultrasonography of the abdomen were normal. Postoperatively the patient was fed through a Ryles tube for 10 days, after which oral feedings were started. Albendazole was given in 400-mg doses twice daily for 12 weeks. Postoperative MR images at the 6-month follow up showed no recurrence of the cysts (Fig. 4); at the 1-year follow up, the patient remained asymptomatic and the cervical reconstruction appeared to be stable.

Discussion

Echinococcosis, or hydatid disease, is caused by the larval form of the tapeworms of the *Echinococcus* genus. This genus includes *Echinococcus granulosus*, *Echinococcus multilocularis*, *Echinococcus vogeli*, and *Echinococcus oligarthus*. The life cycle of this worm involves two hosts: the definitive hosts are dogs, wolves, jackals, and foxes, whereas the intermediate hosts are humans, cattle, sheep, goats, and horses. The adult tapeworm lives in the intestines of the definitive host and discharges eggs into the feces, which are ingested by the intermediate host. Oncospheres released by the eggs are swallowed by humans, invade the duodenum, and enter the portal circulation. From the portal circulation, the oncospheres can lodge in the liver, lungs, and other organs and develop into hydatid cysts. The hydatid cysts contain numerous scolices, each of which is capable of developing into an adult tapeworm. The hydatid cyst is made up of an ectocyst (host tissue response), endocyst, and inner germinal layer. Spinal involvement occurs either through vertebral–portal venous anastomosis or by direct extension of a hepatic or pulmonary focus.

Primary hydatid disease involving the spine is rare, even in regions endemic to this disease, and constitutes only 1% of the disease spectrum.

Moreover,
In one study of skeletal hydatid disease, CT scans in cases of hydatid disease with recurrent and residual disease have included those that detect serum antibodies or circulating antigens. The passive hemagglutination test and enzyme-linked immunosorbent assay are sensitive tests that are used for primary serological screening. However, these tests have low specificity for other cestode infections. A specific immunodiagnostic approach to echinococcosis relies on the demonstration of serum antibodies precipitating an antigen called antigen 5. Monoclonal antibodies have been used against different parasitic antigens to detect circulating antigens in patients’ sera or native proteins in biopsy specimens. Such monoclonal antibodies have been mainly directed against two major antigens—antigen 5 and antigen B. Using these antibodies to detect circulating antigens has been proposed as an early marker of hydatid infection; it can also be used at postsurgical follow up to monitor cyst growth dynamics and activity. Techniques such as DNA hybridization and polymerase chain reaction have been used in the molecular diagnosis of hydatid disease.

Neuroimaging techniques for detecting spinal hydatid disease have included plain radiographs, CT scans, and MR imaging. Lacunar osteolysis is the most characteristic indicator of hydatid osteopathy seen on plain radiographs. In one study of skeletal hydatid disease, CT scans showed well-defined single or multiple nonenhancing cystic lesions. The cystic lesions of hydatid disease can have a honeycomb appearance; there can be associated pathological fractures, bone expansion, cortical thinning, cortical destruction, bone sclerosis, and soft-tissue extension. Typical features of cystic echinococcosis in parenchymal organs such as daughter cysts, calcifications, and germinal membrane detachment are not observed in skeletal hydatid cysts.

Magnetic resonance imaging is the most valuable imaging modality in spinal hydatid disease, for both preoperative planning and postoperative follow up. Magnetic resonance imaging in spinal hydatid disease reveals cysts with thin regular walls without septae and no debris in the lumen. On T2-weighted images, the cyst fluid has a low intensity and the cyst walls are hyperintense, but the cyst fluid is hyperintense on T1-weighted images. It may be possible to distinguish between viable and dead cysts with MR imaging because dying cysts show a decrease in hyperintensity on T1-weighted images and an increase in hypointensity from the collapsed cyst wall. Extralad cysts are always multiple and involve the bone, whereas intradural cysts may be single or multiple. Computed tomography and MR imaging are complimentary when used in the diagnosis of hydatid disease; CT demonstrates the bone changes better, whereas MR imaging is superior for demonstrating neural compression. Braithwaite and Lees have classified spinal hydatidosis into five types: intramedullary, extradural examedullary, extradural intraspinal, vertebral, and paraspinal. The differential diagnosis of vertebral body echinococcosis includes Pott disease, syphilis, bacterial or fungal osteomyelitis, osseous angiomia, chondroma, sarcoma, and hyperparathyroidism.

The primary treatment of spinal hydatid disease is excision of the cysts with concurrent steroid and antihelminthic administration, which is continued in the postoperative period. The drugs commonly used in the treatment of echinococcosis include albendazole, mebendazole, flubendazole, and praziquantel. Immunological tests used in the diagnosis of hydatid disease include spillage of the cyst contents, which can lead to anaphylaxis and secondary dissemination. Albendazole has been used in spinal hydatid disease for periods ranging from 12 weeks to 1 year. There is one report in which albendazole was used intermittently for 9 years. In cases of hydatid disease with spinal instability, spinal instrumentation has been successfully performed. Recurrent and residual disease have been treated with either repeated surgery or medical management of the illness.

One of the dangers during surgery for spinal hydatid disease includes spillage of the cyst contents, which can lead to anaphylaxis and secondary dissemination. This
Hydatid cysts at the craniovertebral junction

hazard can be prevented by using steroids during the procedure, and dissemination can be prevented by injecting the cyst with scolicidal agents prior to excision; these scolicidal agents include formalin, cetrimide, ethanol, hypertonic saline, silver nitrate, glycerin, phenol, ether, dilute betadine, and sodium hypochlorite.5,7,17,22,23

The prognosis of patients with spinal hydatid disease has been varied, ranging from complete eradication of the disease with a resultant cure, to cases with multiple recurrences, systemic dissemination, and death. Recurrence rates from 18 to 100% in large patient series have been reported.1,5,9,19,25 Investigators in one study suggest that the mean life expectancy after spinal involvement is 5 years.14 In view of these features, spinal hydatidosis, although an infection, has been likened to a malignant process. There are encouraging reports of radical surgery involving aggressive removal of the diseased vertebral body with good long-term outcomes.13,15

There has been one report of a case of multiple hydatid cysts of the upper cervical spine and skull base that was diagnosed with a fine needle aspiration biopsy procedure and managed medically with albendazole.7 This patient did not have any neurological deficits or spinal instability. Spektor et al.23 reported a case of hydatid cysts of the upper cervical spine with severe cord compression and neurological deficit that was successfully treated using percutaneous CT-guided aspiration via a lateral cervical puncture and irrigation with hypertonic saline, followed by albendazole therapy. Bozbuga et al.4 reported a case of CVJ hydatidosis in a patient who presented with neck pain and was managed with a posterior stabilization procedure followed by transoral excision of the cysts. The patient received six cycles of albendazole postoperatively and had a good outcome.

Our patient had neurological deficits, significant pain on neck movement, and extensive disease, which led us to attempt a radical excision of all the cysts via anterior and posterior approaches followed by a stabilization procedure. She received albendazole for 3 months, and this treatment along with aggressive surgery resulted in a good outcome. We believe that aggressive surgery followed by chemotherapy offers a chance for disease-free survival with this illness, even in cases of extensive spinal hydatid disease.

References


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