Management of a Rare and Dangerous Infectious Lesion: Hydatid Cyst Disease of the Odontoid Process

Sukru Caglar, Melih Bozkurt, Gokmen Kahilogullari, Mevci Ozdemir

Department of Neurological Surgery, Ankara University, Faculty of Medicine, Ankara, Turkey

ABSTRACT

BACKGROUND: We aimed to demonstrate the multidisciplinary management of hydatid disease of the upper cervical spine and the role of albendazole in the treatment protocol.

CASE REPORT: The patient was evaluated clinically and with laboratory investigations using the latest technological modalities. Endoscopic surgery was performed and the diagnosis was confirmed by histopathologic examination, and the patient received long-term albendazole therapy. After medical treatment, the patient improved temporarily; however, over time, the disease showed exacerbation. Endoscopic endonasal decompressive surgery and posterior stabilization were performed.

CONCLUSIONS: Spinal hydatidosis always necessitates surgical decompression. Albendazole alone or in combination with praziquantel should be given to prevent recurrence.

KEY WORDS: Albendazole, Echinococcus granulosus, Hydatidosis, Infection, Spinal cysts, Tetraplegia, Tuberculosis

Hydatid cyst disease is rare in developed countries, but remains a considerable health problem in endemic areas such as Turkey. Involvement of bones is also rare, occurring in only 0.5 to 2% of cases. Spinal hydatidosis exclusively affects the thoracic spinal column. To our knowledge, there have been only three cases reported in the literature involving the craniocervical region (2,4,14). In the present case, we report a case of cyst hydatid disease involving the craniocervical junction and its multidisciplinary management, including endonasal endoscopic decompression and removal in addition to medical treatment with albendazole and posterior fixation surgery.

CASE REPORT

A 48-year-old male was referred to our clinic with the complaints of neck pain and left upper arm pain and weakness. Six months before admission to our hospital, vertebral tuberculosis was suspected in another center, and rifampicin, isoniazid and pyrazinamide treatment had been applied; neither decompressive surgery nor biopsy had been performed. No diagnosis was revealed according to serological and microbiological studies. His medical history revealed an anterior cervical disc operation at the C5-6 level.

Neurological examination revealed 3/5 monoparesis and sensation deficit in the left C6 dermatome. Hoffman and Babinski signs were positive on the left side. Cervical magnetic resonance imaging (MRI) was performed, and a multilobular cystic, non-septated lesion was found invading the odontoid process. The walls of the cyst were very thin and regular, and the cyst was hypointense on T1-weighted (W) images and hyperintense on T2W images (Figure 1A). Craniocervical junction computed tomography (CT) revealed severe destruction of the C1 anterior arcus, odontoid process and C2 vertebral bodies (Figure 1A). Biopsy was performed by the endoscopic

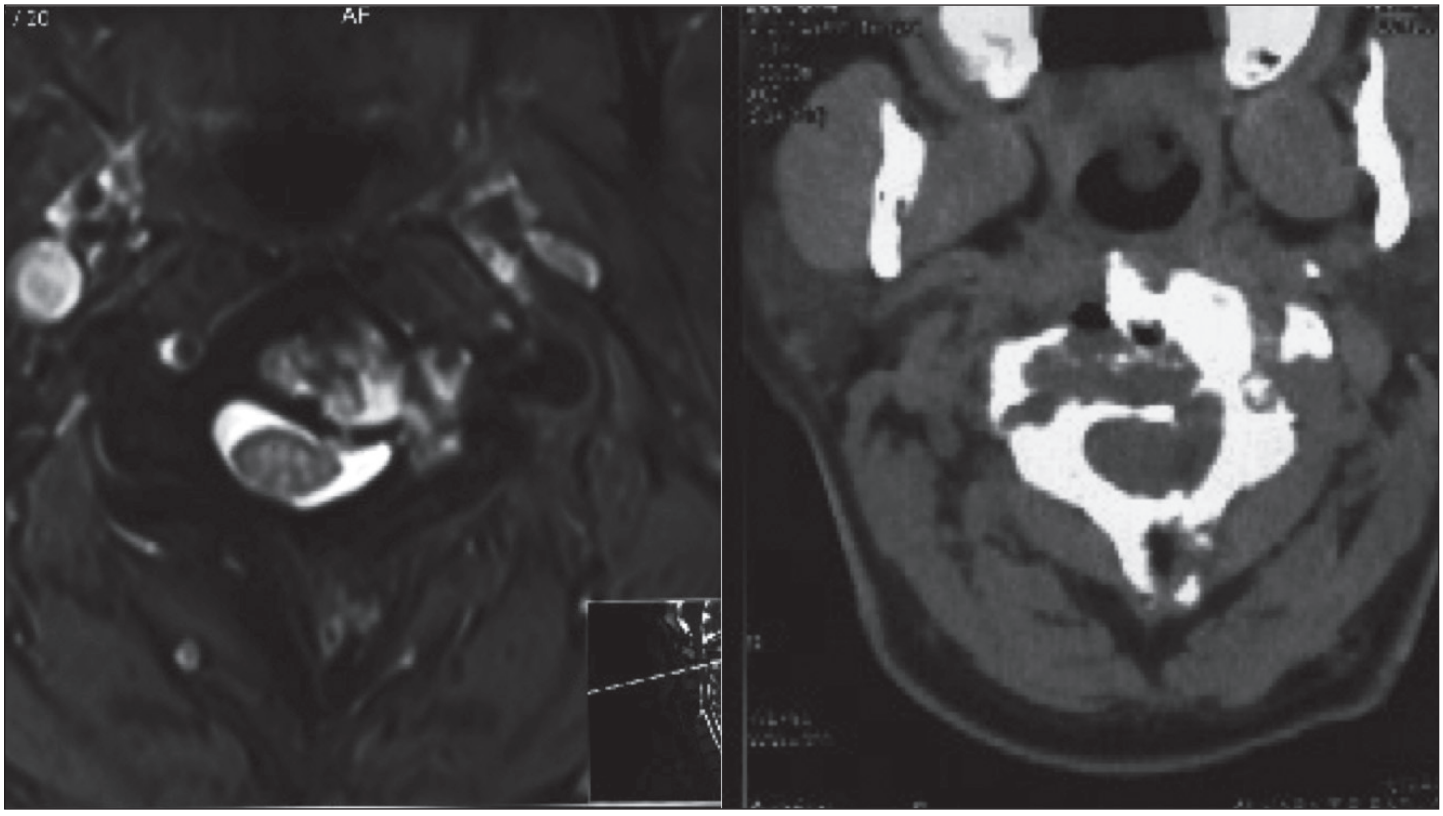

Figure 1: *Axial MR image (A) and CT image showing multilobular cyst invading the odontoid process.*

endonasal route. The pathological study was evaluated as "hydatid cyst". Posterior decompression and fixation were planned but refused by the patient. Albendazole treatment (2 x 400 mg/day) was started after consultation to the Infectious Diseases Department. The patient's liver functions were monitored monthly. After four weeks of treatment, the patient's complaints worsened, and 2/5 monoparesis of the left arm was found. On his repeat MRI, progression of the cyst dimensions was detected. However, the patient again refused the operation, so continuation of albendazole treatment at the same dose was decided on. After 3 weeks, the patient's neurological examination revealed 3/5 monoparesis and his complaints had decreased. At the one-year follow-up, the patient's complaints had worsened again, and the neurological examination revealed 1/5 paresis of his left arm and 3/5 paresis of his left leg. Endoscopic-endonasal excision of the cyst, odontoidectomy, C1-2 laminectomy, and posterior occiput-cervical stabilization were performed (Figure 2). Postoperative neurological examination revealed 4/5 monoparesis of his left arm and leg.

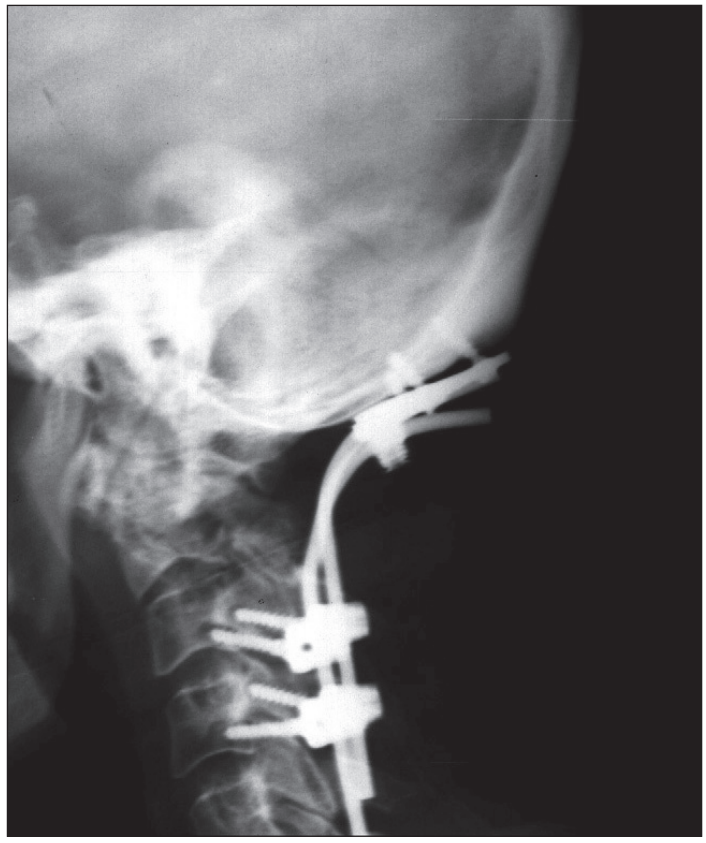

Figure 2: *Postoperative plain x-ray showing the implant.*

DISCUSSION

Hydatidosis is caused by Echinococcus granulosus and less commonly by Echinococcus multilocularis. The most common sites of infection are the liver and lungs. The spinal form was first described by Churrier in 1807, and the first surgery was performed by Reydellet in 1819 (10). Hydatid cyst disease usually extends to the spine by the direct extension from pulmonary, abdominal or pelvic disease (5, 6). Therefore, the cysts are located most commonly at the thoracic (52%), followed by lumbar (37%) and then the sacral levels; cervical localization is quite rare. The infestation persistently erodes the spinal column (11). Destruction of the bones causes mechanical instability and secondary neurological damage due to pathological fractures (7).

There are no specific signs or symptoms in spinal hydatid disease. A long-lasting back pain and/or signs of spinal cord compression are the frequent manifestations. In the presence of suggestive radiological findings, the diagnosis should not be excluded; even a negative serology has been reported. However, the sensitivity falls suddenly to 25%-56% in extrahepatic disease, limiting the use of radiological tools in the diagnosis of primary bone disease (1).

CT usually shows irregular erosions of the cancellous bone and can be used in the diagnosis of bone involvement. On the other hand, MRI is the most sensitive diagnostic method for determination of the extent of the disease. MR images reveal 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 those of cerebrospinal fluid (3, 13). The differential diagnosis should include tuberculosis, pyogenic infections, fibrous dysplasia, enchondroma, metastatic malignancy, multiple myeloma, and giant cell tumor (7).

The goal of surgery is to remove all of the cysts in the early course of the disease. Reported procedures involve posterior approaches with a laminectomy at the cervical region, combined with antihelminthic therapy (8). After excisional surgery, the surgical area needs to be irrigated with hypertonic saline (6). Surgery entails trying to excise cyst aggressively and without rupture and spillage. Not sticking to this plan will lead to definite recurrence. Injection of hypertonic saline and isolation of the surgical wound with chlorhexidine packs are important adjuncts; however, following a cyst rupture, the usefulness of these scolicidal agents is doubtful and recurrences are common. Rupture of a cyst with leakage of the content may induce a group of hypersensitivity reactions such as pruritus, urticaria, edema, dyspnea, asthma, vomiting, diarrhea, abdominal colic pain, and even anaphylactic shock (7, 9, 15).

Albendazole is the first-choice antihelminthic drug in the treatment of cyst hydatid. Its use is offered, in cycles, for 3 months to 1 year after surgery (15). The drug may alter hepatic functions, and hematological parameters should thus be monitored in a cyclic manner. Some authors have reported the effectiveness of albendazole treatment alone for surgically inoperable lesions. However, surgery is still the "gold standard" in the treatment of spinal cyst hydatid. On the other hand, some authors advocate that postoperative albendazole therapy only retards recurrence (6, 8, 12, 15). It is reported that the drug praziquantel is also effective against echinococci alone or in combination with albendazole. Regardless of the controversial data, Lam et al. reported a non-operated case of spinal hydatidosis cured with a combination of albendazole and praziquantel who survived for 34 years (6). The patient presented herein was treated with 800 mg (2 divided doses per day) of albendazole alone, and clinical and radiological findings recovered 6 months after the beginning of treatment. The patient remained in good condition 1 year after the treatment.

CONCLUSION

Despite the introduction of modern surgical and pharmacological therapy, the management and cure of hydatid disease of the upper cervical spine continue to be difficult, and the disease is highly prone to recurrence (30%-100%), requiring extensive and repeated spinal surgeries with high rates of complications and significant long-term morbidity and mortality. Operative mortality rates up to 15% are noted in the literature and are known to increase with subsequent operations. Paraplegia due to recurrent disease is reported to be as high as 45%. The prognosis is poor, and has been compared with that of spinal malignancy (7, 8, 15). We have reported herein a rare case of spinal column involvement. In addition to the unusual localization of this tumor in the spinal column, the other significant feature was the treatment

modality. The two significant points of this case are that spinal hydatidosis always requires surgical decompression and that albendazole alone or in combination with praziquantel should be given to prevent recurrence.

REFERENCES

1. Baykaner MK, Dogulu F, Ozturk G, Edali N, Tali T: A viable residual spinal hydatid cyst cured with albendazole. J Neurosurg Spine 93:142–144, 2000
2. Bozbuga M, Celikofu E, Boran OB: Hydatid cyst of the craniocervical junction: case report. Neurosurgery 57:193-195, 2005
3. Doganay S, Kantarci M: Role of conventional and diffusion-weighted magnetic resonance imaging of spinal treatment protocol for hydatid disease. J Spinal Cord Med 32(5):574–577, 2009
4. El Kohen A, Benjelloun A, El Quessar A, Derraz S, Lazrak A, Jazouli N, Kzadri M: Multiple hydatid cysts of the neck, the nasopharynx and the skull base revealing cervical vertebral hydatid disease. Int J Pediatr Otorhinolaryngol 67:655–662, 2003
5. Kalkan E, Cengiz SL, Cicek O, Erdi F, Baysefer A: Primary spinal intradural extramedullary hydatid cyst in a child. J Spinal Cord Med 30:297–300, 2007
6. Lam KS, Faraj A, Mulholland RC, Finch RG: Medical decompression of vertebral hydatidosis. Spine 22:2050-2055, 1997
7. Ozdemir HM, Ogun TC, Tashas B: A lasting solution is hard to achieve in primary hydatid disease of the spine: long-term results and outcome. Spine 29:932–937, 2004
8. Ozer AF, Ozek MM, Pamir MN, Erzen C: Magnetic resonance imaging in the diagnosis of spinal hydatid cyst disease. Case report. Paraplegia 31:338–340, 1993
9. Pamir MN, Akalan N, O Ozgen T, Erbengi A: Spinal hydatid cysts. Surg Neurol 21:54-57, 1984
10. Pamir MN, Ozduman K, Elmaci I: Spinal hydatid disease. Spinal Cord 40:153–160, 2002
11. Prabhakar MM, Acharya AJ, Modi DR, Jadav B: Spinal hydatid disease: a case series. J Spinal Cord Med 28:426–431, 2005
12. Schnepper G, Johnson WD: Recurrent spinal hydatidosis in North America. Case report and review of the literature. Neurosurg Focus 17:1–6, 2004
13. Secer HI, Anik I, Celik E, Daneyemez MK, Gonul E: Spinal hydatid cyst mimicking arachnoid cyst on magnetic resonance imaging. J Spinal Cord Med 31:106–108, 2008
14. Semlali S, Nassar I, Cisse A, El-Gueddari FZ, Imani F: Cervical hydatid disease with retropharyngeal involvement. A case report. J Radiol 85:51–53, 2004
15. Sucu K, Zileli M: Management of spinal infections [in Turkish], in Zileli M, Ozer F (eds): Spinal Cord and Spine Surgery [in Turkish]. Izmir, Meta Basım, 2002:1145–1173

Manuscript submitted April 13, 2012.
Accepted June 04, 2012.

Address correspondence to: Y. Sukru Caglar, Department of Neurological Surgery, University of Ankara Ibni Sina Hospital, Ankara, Turkey

Phone: +90 505 319 13 30
Fax: +90 312 426 23 11

email: 23sukrucaglar@yahoo.com