

Orbital hydatid cyst

Case report

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Hydatid cysts rarely appear in the orbital cavity without the involvement of the other organs. Most of them are situated in the superolateral and superomedial angles of the orbit. Inferiorly located cysts are very uncommon. The authors report a case of a primary hydatid cyst of the orbit with inferolateral localization. The cyst was removed surgically via a frontotemporoorbitozygomatic approach combination with puncture-aspiration-injection-reaspiration technique. This case was considered as a primary infection, because there was no previous history of hydatid disease and no findings of liver and lung cysts on radiological examination. Physicians should include orbital hydatid cyst in the differential diagnosis of unilateral proptosis. To avoid complications that might occur during surgery, the cyst can be easily removed using the combination technique detailed in this report.
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KEY WORDS • primary • orbital • hydatid cyst • congenital •
inferolateral localization • frontotemporoorbitozygomatic • scolicides

HYDATID disease is caused by the larval stage of *Echinococcus* tapeworms. The disease is endemic in societies in contact with livestock. Hydatidosis may involve almost every organ or tissue via the portal and systemic circulations in humans. The liver and lungs, followed by the CNS, are the most frequent sites of hydatid cysts, but orbital involvement is exceptionally rare, even in highly endemic regions. Hydatid infestation of the orbit comprises far less than 1% of the total incidence.^{11,19,20} The most appropriate treatment of an orbital hydatid cyst is surgical removal. However, dissemination of the disease due to cyst rupture during surgery is the most feared complication. Different surgical approaches have been previously described for removal of the orbital hydatid cyst, with the transcranial approach used the most frequently. The size of the cyst, its anatomical location, the patient's general medical condition, and the surgeon's familiarity are the key elements for selection of the most suitable approach.⁴ Careful preoperative radiological evaluation coupled with clinical findings may allow differentiation of the hydatid cyst from other orbital space-occupying lesions. Nevertheless, this differentiation is difficult when based only on the radiological appearance of a hydatid cyst when it is solely located in the orbital cavity. Therefore, intraoperative aspiration of the cystic contents is recommended. However, puncture of the cyst can lead to some spillage of the contents into the operative field and dissemination of the disease; therefore, in situ irrigation of the cystic cavity

with scolicides should be performed during surgery. This combined method is mostly preventative for rupture of the cyst and dissemination of the disease. In addition to surgical treatment, systemic use of antiparasitic drugs is recommended for both pre- and postoperative periods.^{14,17,19}

Case Report

History and Examination. This 6-year-old previously healthy girl, who had a history of close contact with dogs and sheep, presented with a history of gradual vision loss and painless proptosis on her left eye for 3 months. Ophthalmological examination revealed conjunctival chemosis and mild proptosis. Her best-corrected visual acuity was 4/10 in her left eye and 10/10 in her right eye. Fundoscopic examination revealed a blurred optic disc. There were no other abnormalities in her general examination. Her blood count and routine laboratory investigations were normal. Axial, sagittal, and coronal MR images revealed a lesion with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, which displaced the optic nerve superomedially and the globe anteriorly, with fine peripheral rim enhancement of its fibrous capsule after contrast medium administration (Fig. 1). Serological tests were performed to exclude parasitic disease, but all were negative. Preoperatively, with the support of radiological findings, orbital hydatid cyst was suspected.

Orbital hydatid cyst

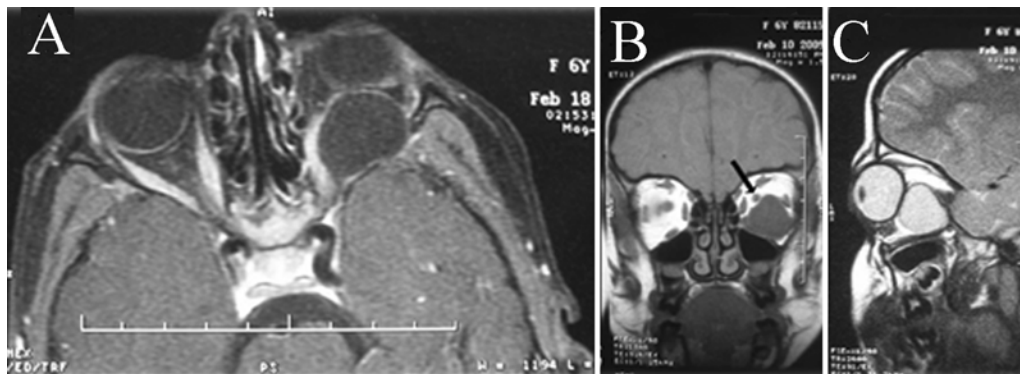


Fig. 1. Preoperative MR imaging of the patient. **A:** Axial T1-weighted image of the left orbita with Gd injection demonstrating globe deformation and proptosis of the orbit due to a retroorbital huge cystic lesion with fine peripheral rim enhancement. **B:** Coronal T1-weighted image showing an inferolaterally localized cystic lesion with low signal intensity displacing the optic nerve superomedially (arrow). **C:** Sagittal T2-weighted image showing a cystic lesion with high signal intensity displacing the globe anteriorly.

Operation and Postoperative Course. During surgery a left-sided frontotemporozygomatic craniotomy and orbitotomy was performed. A unilocular cyst was found in the inferolateral aspect of the orbital cavity. A sharp 27-gauge needle with a syringe was gently introduced into the cyst cavity, and a transparent clear fluid was aspirated completely, which was characteristic for a hydatid cyst. Afterward, 5 ml of hypertonic saline solution was lightly injected into the cavity and reaspirated a few minutes later. The inner germinal layer of the cyst (endocyst) was collapsed, so that the outer fibrous wall was easily tented with forceps and removed completely. No additional neurological deficit was obtained after surgery. Her left-side proptosis was completely resolved within 2 weeks postoperatively. Histopathological examination of the cyst fluid confirmed the diagnosis of hydatid disease. Afterward, chest and abdominal CT scans of the patient were obtained to rule out systemic involvement, but they showed no evidence of hydatid disease. Therefore, we accepted this case as a primary infection. On postoperative control cranial CT, total excision of the cyst was confirmed (Fig. 2). The patient was discharged from the hospital 2 weeks after the operation with oral albendazole treatment and advised to maintain regular follow-up contact.

Discussion

Hydatid cyst is a worldwide health problem caused by *Echinococcus granulosus*, a tapeworm. This disease is prevalent in Africa, the Middle East, Eastern Europe, and Mediterranean countries, as well as in Turkey. These regions are notable for continuous contact with sheep and cattle.^{11,14,19,20}

Hydatid cyst is more common in the liver (60%–70%) and lungs (20%). Orbital involvement takes place in 1%–2% of all cases. Orbital infestation is more prevalent among young patients with no sex predilection. Patients are often admitted to the hospital with progressive proptosis lasting from 1 week to 2 years.^{11,13,18,19} In our case, it was 3 months. Most patients present with limited ocular motility, decreased visual acuity due to papilledema, chemosis, eyelid edema, and conjunctivitis.^{9,16,18} Chemosis, eyelid edema, and impaired vision with papilledema were

all present in our patient, whereas limited motility was not. The cyst is composed of an outer membrane and inner germinal layer. The inner layer is ultimately fragile.¹¹ Therefore, early diagnosis is important to avoid complications, such as spontaneous cyst rupture causing severe allergic and inflammatory reactions. Prompt surgical excision of the cyst is recommended for prevention of seeding during surgery due to the advanced stage of the disease.^{4,17}

According to clinical and radiological findings, the differential diagnosis of orbital hydatid cyst includes any well-circumscribed, nonpulsatile orbital lesions such as abscess, epidermoid-dermoid cysts, teratoma, and mucocele. Serological tests can be used for the diagnosis of echinococcus, but they usually give negative results as far as primary orbital hydatid disease is concerned. Using only a clinical evaluation with serological findings is not adequate to diagnose an orbital hydatid cyst; radiological findings play an important role for preoperative differential diagno-



Fig. 2. Postoperative axial CT scan showing a decrease of proptosis and soft-tissue changes in the lateral part of the left orbita due to surgery.

sis.^{2,12,20} However, a definitive diagnosis is rarely achieved preoperatively. Orbital ultrasonography is a useful test when a “double wall sign” has been observed.³ Magnetic resonance imaging and CT are both useful techniques for preoperative diagnosis and surgical planning. However, the former is suggested to be more effective due to its capability to provide us with more detailed information about the nature of the cyst and its relationship with the neighboring structures. Computed tomography of the orbital hydatid cyst demonstrates a hypodense, unilocular, well-defined homogenous mass with a hyperdense rim.^{7,21} Magnetic resonance imaging demonstrates a cystic lesion with a low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, with peripheral rim enhancement of the cyst capsule after contrast medium administration, as in our case.^{10,12}

According to previous studies, appropriate treatment of the orbital hydatid cyst is surgical removal.¹ Different surgical approaches can be used for the removal of the orbital hydatid cyst, but the transcranial approach is the most frequently used approach. A parietotemporal microsurgical approach had been recommended by some authors.^{5,8} In our case, the cyst was removed successfully from the retroorbital region via a left-sided frontotemporoorbitozygomatic approach combination with the puncture-aspiration-injection-reaspiration technique. This technique is typically performed in 3 steps: puncture and needle aspiration of the cyst, instillation and indwelling of a scolical solution for a few minutes, and cyst reaspiration. This method produces the collapse of the inner germinal layer, facilitating the total removal of the cyst. Scolicidal solutions used in the puncture-aspiration-injection-reaspiration technique included hypertonic saline, absolute alcohol, silver nitrate, and mebendazole. Hypertonic saline solution is the most frequently used scolical solution and more appropriate for neurosurgical procedures because of non-toxic features to the neural tissue.¹⁵ No complications have occurred during surgery when using this solution. It is possible to decompress the orbital cavity and extract the lesion without any retraction of neural structures using the frontotemporoorbitozygomatic approach. This approach allows a well-controlled surgery, especially for inferolaterally located lesions of the orbital cavity.^{4,6}

This case indicates that hydatid cyst should be included in the differential diagnosis of unilateral proptosis in children, especially in regions endemic to this disease. Preoperative radiological examinations should be evaluated in children for differentiation of hydatid cyst from other intraorbital mass lesions and to avoid complications from advanced disease. Surgical excision of the orbital hydatid cyst in combination with the puncture-aspiration-injection-reaspiration technique can be performed safely and easily by experienced neurosurgeons.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Kahveci. Acquisition of data: Sanli. Drafting the article: Kahveci, Güreç, Sekerci. Critically revising the article: all authors. Reviewed submit-

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