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CASE REPORT

Isolated primary orbital hydatid cyst: Two cases

Melis Palamar,[1] Derya Dirim Erdogan,[2] Taner Akalin,[3] Oguz Resat Sipahi[4]

[1]Department of Ophthalmology, Ege University Faculty of Medicine, Izmir, Turkey
[2]Department of Parasitology, Ege University Faculty of Medicine, Izmir, Turkey
[3]Department of Pathology, Ege University Faculty of Medicine, Izmir, Turkey
[4]Department of Infectious Diseases, Ege University Faculty of Medicine, Izmir, Turkey

Abstract

Hydatid cyst is a rare parasitic disease caused by *Echinococcus granulosus* or *Echinococcus alveolaris* tapeworm. The most common sites that are affected are liver, lung, and central nervous system. Other rarely affected sites are orbit and bone. Herein, two cases of isolated primary orbital hydatid cysts that were surgically managed are presented.

Keywords: Cyst hydatid; *Echinococcus alveolaris*; *Echinococcus granulosus*; orbit; ptosis.

Hydatid cyst is a rare parasitic disease caused by *Echinococcus granulosus* or *Echinococcus alveolaris* tapeworm. Hydatid cyst is one of the most common zoonotic diseases worldwide and a major public health problem, especially in endemic places. It has a predilection of regions where sheep raising is common.[1] The definitive hosts are commonly dogs and other Carnivora.[1] Hydatid cysts develop in sheep and cows, while the larval stage takes place in humans.[2] The most common sites that are affected are liver, lung, and central nervous system. Other rarely affected sites are orbit and bone. Incidence of orbital hydatid cyst is <1% in a year in the literature.[1] Isolated primary orbital hydatid cyst without involvement of any other organ is very rare. Herein, two cases of isolated primary orbital hydatid cysts that were surgically managed are presented.

Case Report

Case 1– A 10-year-old girl with a slowly growing mass lesion for 2 months in the inferior orbit was admitted to the clinic. She had no particular history of trauma. The well-limited fluctuating lesion was causing superior displacement of the eye resulting with diplopia (Fig. 1a). The visual acuity was 20/20, and the eye and eye movements were normal. In blood count, mild monocytosis was present. Magnetic resonance imaging (MRI) of the orbit demonstrated a well-limited cystic lesion in the inferior orbit originating from the ventral region of the inferior rectus (Fig. 1b). A subciliary orbitotomy was performed to excise the whole cyst. However, the cyst ruptured during excision. The surrounding area was rinsed with hypertonic saline solution and betadine. The histopathologic examination of the

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Correspondence: Melis Palamar, M.D. Department of Ophthalmology, Ege University Faculty of Medicine, Izmir, Turkey
Phone: +90 232 390 37 88 **E-mail:** melispalamar@gmail.com
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lesion revealed hydatid cyst (Fig. 1c and d). Five different serum samples of the patient were tested by in-house enzyme-linked immunosorbent assay (ELISA) immunoglobulin (Ig) G[3] and indirect hemagglutination (IHA)[4] to detect antibodies against *E. granulosus*, in the 6-month period after the operation. Specific anti-*E. granulosus* antibodies were detected 1/80 positive with ELISA IgG and 1/160 positive with IHA on the 45th day after the operation. While other serum samples were detected negative by anti-*E. granulosus* ELISA IgG and IHA, only one serum sample was detected as 1/80 positive by IHA, on the 80th day after operation. The patient was consulted to Infectious Diseases Clinic and was scanned for other foci elsewhere. She was found to have no other involvement. However, albendazole treatment was initiated and continued for 6 months. She experienced no recurrence for 60 months.

Case 2– A 29-year-old male experiencing left upper eyelid swelling and ptosis with accompanying mild conjunctival hyperemia and foreign body sensation for 5 months was referred to the clinic (Fig. 2a). The symptoms were unresponsive to systemic antibiotics and non-steroidal anti-inflammatory drugs. No trauma history was present. The visual acuity was 20/20. Eye movements were normal. When the upper eyelid was everted, a granuloma-like lesion was evident in the superior orbit (Fig. 2b). In blood count, lymphomonocytosis was present. Orbital MRI demonstrated an abscess focus located in the upper eyelid with neighboring infection signs (Fig. 2c). A superior conjunctival orbitotomy was performed to totally excise the mass. The histopathologic evaluation revealed an *E. alveolaris*-related inflammatory lesion (Fig. 2d and e). The serum sample of the patient was tested to detect antibodies against for *E. granulosus* and *Echinococcus multilocularis* on the 5th day after operation. Specific anti-*E. granulosus* antibodies were detected 1/160 positive by ELISA IgG and negative by IHA and anti-*E. multilocularis* IgG was determined negative by

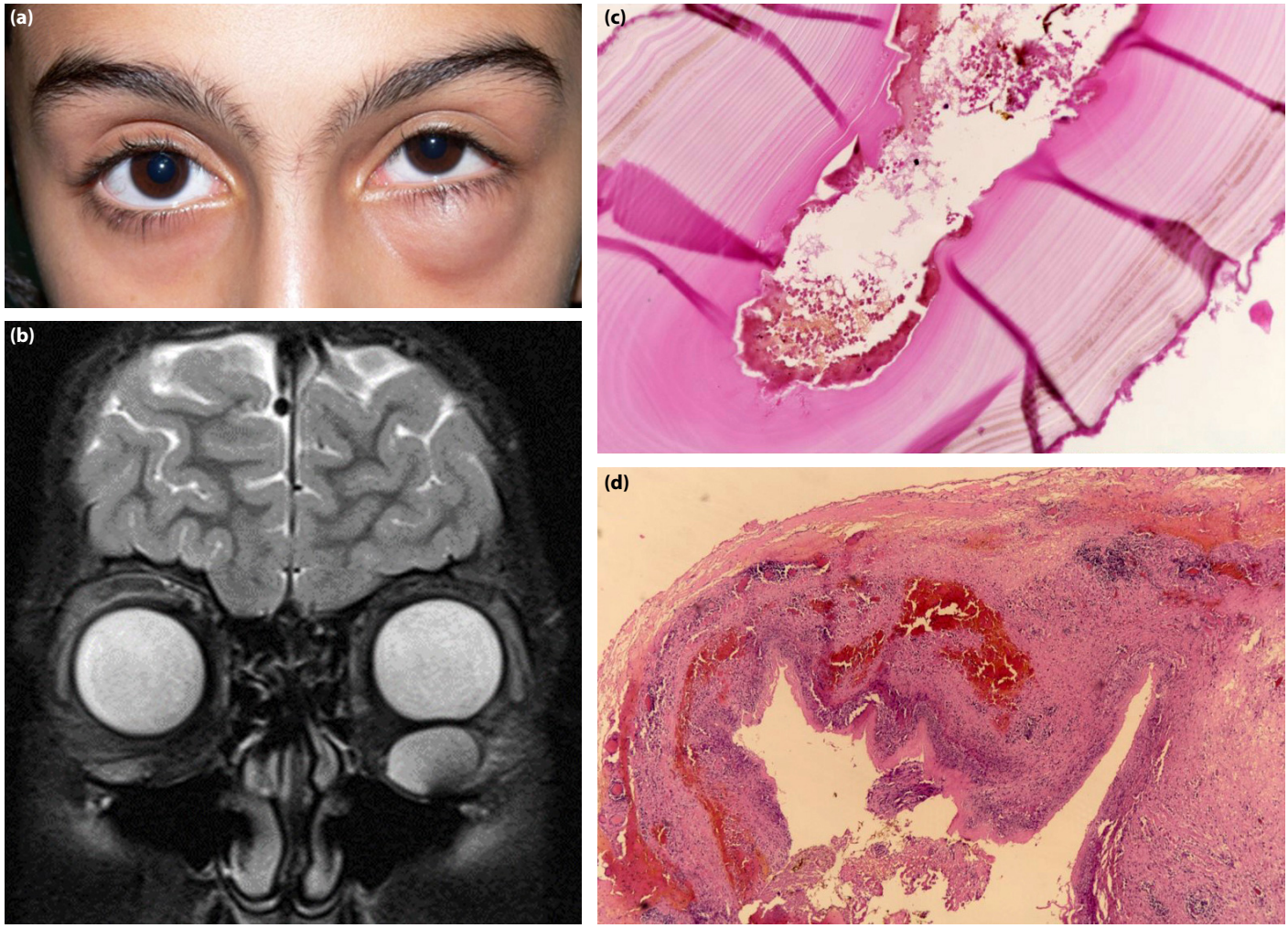

Fig. 1. **(a)** Appearance of the 10-year-old female at presentation. **(b)** Cranio-orbital magnetic resonance imaging demonstrating the infraorbital cystic lesion. **(c and d)** Histopathologic examination revealed fibrous laminar (cuticular) membrane **(c)** and surrounded by granulation tissue characterized with histiocytes, inflammatory cells, and fibrous tissue **(d)** compatible with hydatid cyst.

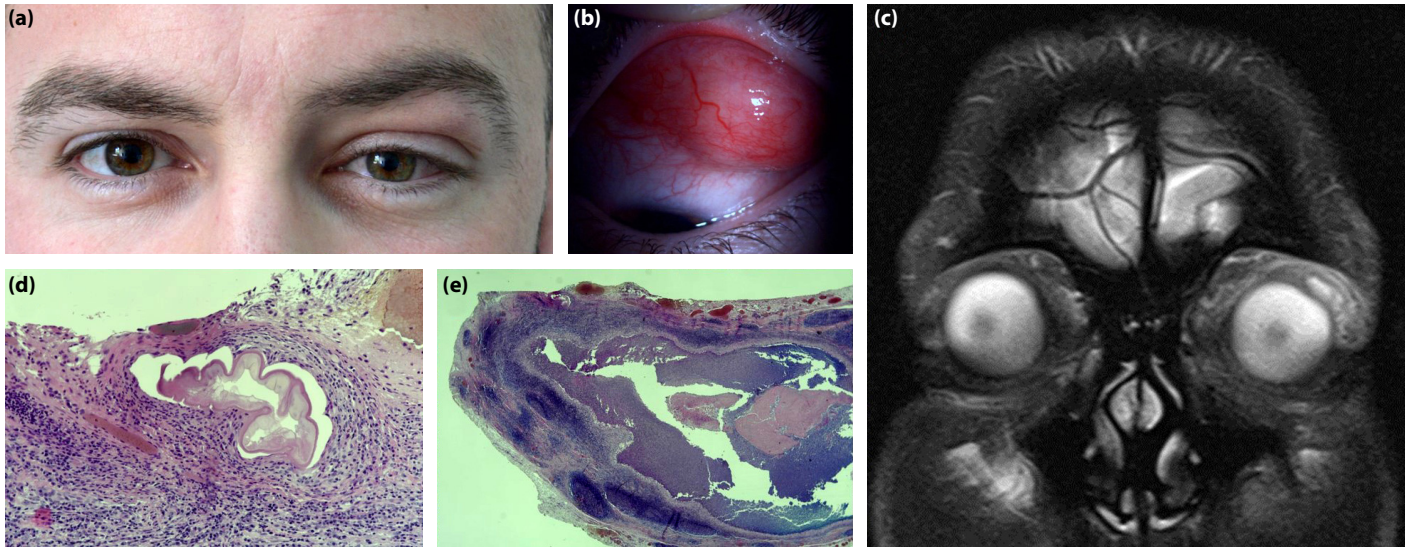

Fig. 2. **(a)** Left upper eyelid swelling and ptosis in the 29-year-old male. **(b)** A superior forniceal lesion with accompanying mild hyperemia is evident. **(c)** Cranio-orbital magnetic resonance imaging showing a heterogenic hyperintense focal, abscess-like lesion in the left superior eyelid. **(d and e)** Histopathologic examination revealed laminar membrane (d) surrounded by granulomatous reaction and heavily inflammatory cells around **(e)** compatible with hydatid cyst.

in-house immunoblotting.[5] The patient was consulted to Infectious Diseases Clinic and was scanned for other foci elsewhere which revealed no other involvement. Albendazole treatment was initiated and continued for 6 months. No recurrence or other site of involvement occurred for 66 months.

Discussion

The prevalence of hydatid cyst is high in Africa, East Europe, Mediterranean, and the Middle East regions where people frequently contact with sheep and cattle.[1] Orbital hydatid cyst is a rare entity that usually is observed in young population, especially in children.[1] It has no gender predilection.[1] Orbital hydatid cyst is generally unilateral with the left side affected more than the right.[6,7] The most common locations of orbital hydatid cyst are intraconal, superomedial, or superolateral quadrants. Both of our cases young, one being a child, and both had left-sided lesions. One of our cases was infraorbital and one was located within the eyelid, both of which are atypical locations. None of the cases were in contact with sheep or cattle or even dogs.

The slow type of growth in hydatid cysts results in capsular fibrosis formation surrounding the two-layered wall of cyst and clear antigenic fluid.[8] These two layers are the outer laminated ectocyst as a barrier layer in avoiding tissue invasion and the inner germinal layer that contains the daughter cysts. The daughter cysts create tape worm by producing scolices.[8] For this reason, cyst removal without its wall damage prevents spread of scolices and possible recurrence.[8] However, the complete removal of the cyst without any rupture is not very easy.

The most common clinical manifestation of hydatid cyst is slowly progressive unilateral proptosis with or without pain.[2] Keratoconjunctivitis, eyelid edema, chemosis, restricted eye movements, and even visual loss might accompany the cystic mass.[2,6,7] Both of our cases were slowly progressive. However, the clinical manifestations were related to the localization of the mass. The first case in whom the cyst was infraorbital besides the swelling in the affected area, diplopia due to superior displacement of the globe was evident. In the second case, the mass within the eyelid caused both a swollen eyelid and ptosis. Moreover, the mass lesion located in the eyelid caused foreign body sensation and mild reactive conjunctival hyperemia. The atypical eyelid localization probably ended up with an abscess instead of a cyst formation. For this reason, MRI could not detect a cyst but an abscess. Although the lacrimal gland was near the hydatid abscess, it was not involved. The slow progressing, low inflammatory nature of the hydatid abscess caused a delay for surgical excision.

The differential diagnosis of an orbital hydatid cyst includes lesions such as abscess, epidermoid or dermoid cysts, hematocele, teratoma, encephalocele, and mucocele.[2] Although, clinical evaluation along with radiological imaging techniques usually is sufficient for diagnosis, some cases still need histopathologic confirmation. For example, in the second case presented here, the abscess formation did not recall hydatid cyst at all.

The treatment of hydatid cysts is mainly surgical. However, pre- and post-operative albendazole courses of 1 month and 2 weeks of praziquantel should be considered to sterilize the cyst, decrease the risk of anaphylaxis, reduce the tension in the cyst wall, and reduce the post-operative recurrence rate.[8]

Surgical treatment can be of two types, simple aspiration or total excision.[9] The highly recommended technique is typically performed in three steps: Puncture and needle aspiration of the cyst, instillation and indwelling of a scolicidal solution for a few minutes, and cyst reaspiration puncture, aspiration, injection, and reaspiration (PAIR).[1,10,11] This method produces the collapse of the glistening inner germinal layer, which is gripped with a cryoprobe and completely extracted.[1,10,11] The outer fibrous layer is left behind, eliminating the need for potentially damaging orbital dissection.[11] Scolicidal solutions – which kill the daughter cysts and prevents anaphylactic reactions and further spread of seeding – used in PAIR technique include hypertonic saline, absolute alcohol, silver nitrate, and mebendazole.[1,9] Hypertonic saline solution is the most frequently used scolicidal solution and more appropriate for orbital procedures because of non-toxic features. Following PAIR technique, some authors believe that the cyst might surgically be removed.[12]

The preferred surgical removal approach of the hydatid cyst differs according to the localization and extension of the lesion (fronto-orbital, lateral orbitotomy, transcranial, transconjunctival, inferior orbitotomy, lateral rhinotomy, percutaneous, transmaxillary, and enucleation).[13]

If the cyst accidentally ruptures, *in situ* irrigation with hypertonic saline should be performed.[9,14] It is better to cover the surrounding area of the cyst and exposed optic tissue with hypertonic saline soaked cotton before beginning dissection and cyst removal.[9,14]

The most important factors in prognosis are the localization of the cyst, rupture of the cyst and dissemination of its content, and treatment modality.[6] When an orbital or adnexal cyst is detected – especially in endemic areas – it should be treated as if it is a hydatid cyst. A delay in making the diagnosis and treatment might cause undesired ocular complications, even total visual loss. Surgical excision without causing rupture of the cyst is the sole cure for hydatid cyst. Systemic evaluation for hydatidosis revealed no additional foci in both cases. Post-operative albendazole is known to be effective in ruptured cases, recurrences, and systemic hydatidosis. Both of the presented cases received 6-month course of albendazole and had no relapse during >5-year follow-up period.

Conclusion

As a conclusion, detection of an orbital cyst in endemic areas should recall hydatid cyst. Early diagnosis and prompt management are mandatory for good prognosis. Total removal of the cyst without rupture requires careful dissection of cyst along with the use of scolicidal solutions pre-, intra-, and post-operatively. Moreover, in confirmed hydatid cases, evaluation for accompanying systemic involvements should not be neglected.

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