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CASE REPORT

Primary Orbital Hydatid Cyst

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ABSTRACT

The authors want to highlight the importance of clinical, radiological and histopathological evaluation in unilateral proptosis. A 17-year-old male presented with left progressive nonpulsatile proptosis, lateral gaze diplopia and decreased visual acuity. It was diagnosed as orbital hydatid cyst on CT scan and MRI. The patient was successfully operated with near total recovery of the vision. Radiological investigation showed a circumscribed cystic lesion lateral to orbit separate from the lacrimal gland. Intraoperative cysts and postoperative histopathology confirmed the primary orbital hydatid cyst.

Although rare there is always a possibility of a primary hydatid in patients with unilateral proptosis, restricted eye movements and lateral decreased visual acuity. This is possible even when the Casoni's test is negative. Surgical excision with postoperative albendazole is the effective treatment for the cure of disease.

Keywords: Hydatid cyst, Orbital hydatid cyst, Nonpulsatile proptosis.

INTRODUCTION

Hydatid disease, a parasitic infectious disease caused by larval stage of *Echinococcus granulosus*. It has three host epidemiological chain of sheep, dog and human. Humans are intermediate hosts with hydatid cyst affecting almost all the organs. Liver and lungs are most commonly affected. Here we present a rare case of sero-negative hydatid cyst affecting the orbit diagnosed on the basis of MRI and treated by surgery followed by chemotherapy.

CASE REPORT

A 17-year-old male presented with complaints of progressive painless nonpulsatile proptosis of left eye since 8 months and reduced vision since 5 months.

Ophthalmic examination revealed a slight eccentric non-reducible inferomedial proptosis of left eye and measured 11.2 mm by Hurtel's exophthalmometer with normal anterior segment. Lateral movement of the left eye were restricted and visual acuity reduced to finger counting. There was a nonfluctuant swelling at the lateral canthus of left eye. Fundus examination showed a vertically oval disk with hyperemia and blurred margins (Fig. 1). The blood vessels were stretched and tortuous. There were signs of macular edema with clear media in the posterior segment.

CT scan revealed a nonenhancing, lobulated well circumscribed cystic mass with watery content and separate from the lacrimal gland.

MRI also confirmed the findings showing that the cyst had a hyperdense rim with septa running through the cyst

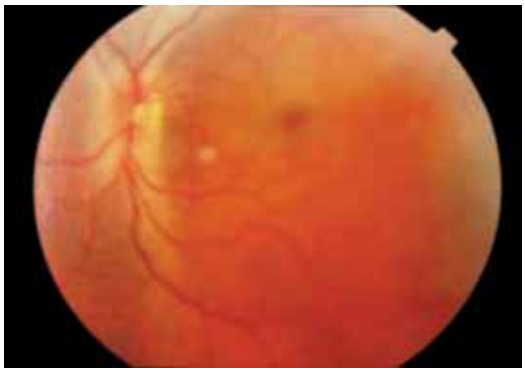

Fig. 1: Fundus picture showing a vertically oval disk with hyperemia and blurred margins

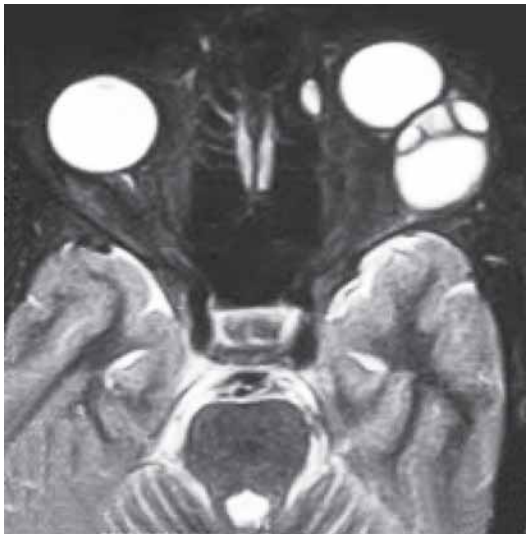

Fig. 2: Cyst with hyperdense rim and septa running through the cyst

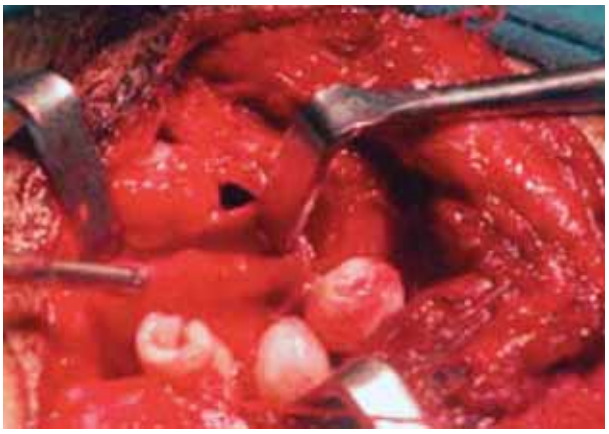

Fig. 3: Multiple daughter cysts within the primary cyst extending to the posterior part of the eyeball

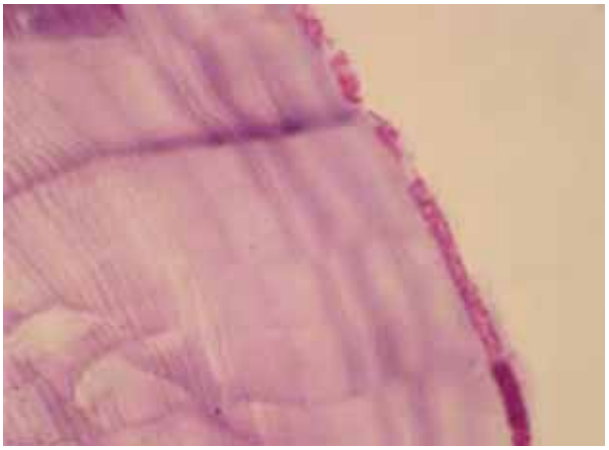

Fig. 4: Histopathology of the encised sample confirmed the diagnosis of hydatid cyst

(Fig. 2). Chest X-ray and abdominal ultrasonography was normal. There was no lymphadenopathy. The routine blood counts were normal, stool and urine examination did not show any abnormality and the Casoni's test was negative.

Patient was taken for cyst excision. Incision was given at the inferolateral orbital margin and the soft tissue showing excessive fibrosis was dissected from the bony orbital wall. The cyst wall was dissected away from the lateral rectus muscle. Multiple daughter cysts (Fig. 3) within the primary cyst extending to the posterior part of the eye ball which were identified and dissected out taking care that they do not rupture. The bony lateral orbital wall was split to remove the cysts intact, and was later plated. The wound was thoroughly washed with hypertonic saline to prevent any recurrence or toxic reaction due to accidental spill of the fluid containing the scolices of the *Echinococcus*. The excised sample was sent for histopathology (Fig. 4) which confirmed the diagnosis of hydatid cyst.

The patient was kept on broad spectrum systemic antibiotics, antiinflammatory drugs and tablet albendazole for next 2 weeks at a dose of 400 mg twice daily. He was followed up for next 6 months and was completed asymptomatic. The papilloedema subsided and the visual acuity recovered to 6/18, although diplopia persisted on left lateral vision.

DISCUSSION

Hydatid disease is caused by the development in man, acting as an intermediate host, of the cystic larval form of *Echinococcus granulosus*. It is one of the commonest parasitic affection in the temporate climate of the world.

Orbital hydatidosis represents a quite rare clinico-pathologic entity. It can either be primary, where cyst or cysts are solely located into the orbital cavity (as in our case) or secondary, when orbital lesions are a part of disseminated multiorgan systemic disease. The usual strains responsible for orbital cysts are *E. granulosus* and *E. multicularis*, although *E. oligarthrus* has been isolated in cases from South and Central America.

Huilgol[1] reported the incidence of orbital cyst from 0.7 to 1 % of all orbital tumors but in highly endemic areas, orbital hydatid cysts have been reported to represent up to 25.8% of all cystic orbital lesions.[2]

Orbital hydatid cysts are solitary lesions in the vast majority of cases, but there are reports in the literature of multiple intraorbital cysts, a phenomenon which occurs in less than 5% of patients with orbital hydatid cysts.[3]

The most commonly presenting clinical symptoms and signs of orbital *Echinococcosis* are: Nonpulsatile, nontender exophthalmos (as in our case), visual disturbances, papilledema, diplopia chemosis, eyelid edema, conjunctivitis and hypopyon.[4] The mean duration of symptoms has been reported to vary between 3 months and 2 years.[5] In our cases, the duration of symptoms was 2 months.

In regards to the anatomic location of the intraorbital hydatid cysts, various orbital locations have been described. It is well accepted that there is a predilection for the superomedial and superolateral orbital areas. Interestingly, Talib, in his report, stated that left-sided lesions were more common than right-sided ones; due to the fact that left, common carotid artery arises directly from the summit of the aortic arch.[6]

These lesions may be diagnosed by various imaging techniques. On CT scan the typical characteristic feature of the 'water-lily' sign of *Echinococcal* cysts in other locations may or may not be present in the orbital cysts. The MRI is superior in the detection of the lesion and the delineation of its relationship to the adjacent ocular structures. Upon MRI examination, the cystic lesion appears low signal on T1W images, high signal on T2W images, capsular and peri-capsular soft tissue enhancement, and the capsule is seen as a hypointense rim surrounding the mass on T2W images.[7]

Because of the integrity of the cyst wall in the orbit, the serologic test results may be inconsistently positive in 50%

of cases compared with 98% positivity in hepatic hydatosis. Casoni's test was found to be negative in our case and it is now widely accepted that none of the currently employed serologic tests can rule out the existence of a solitary orbital hydatid cyst.[8]

The only definite treatment of hydatid cyst is surgical removal. The size of the cyst, its anatomic location, its extension into the cranial cavity, the patient's general medical condition, the extent of the disease and the surgeon's familiarity with each surgical approach decide the most suitable approach. Meticulous microsurgical dissection, continuous irrigation, during dissection and adequate visualization are important for the success. Postoperative adjuvant chemotherapy with albendazole is always recommended.[9] Rupture and spillage of the contents of the cysts leads to secondary dissemination with local recurrence, so postoperative treatment is of great importance. Mebendazole and albendazole have been shown to be effective in such cases.[3]

The outcome of these procedures has been reported excellent in the vast majority of cases; however, in late stage cases with extensive optic atrophy and global compression, enucleation of the globe has been reported.[1]

The early diagnosis of orbital hydatidosis and its prompt surgical management has excellent results in the vast majority of these patients. This makes the importance of its appropriate diagnosis even more crucial. The clinician should always include the diagnosis of hydatid cyst in the differential diagnosis list of orbital mass, especially in children. Increases of world tourism and migration should make the clinician to include this quite rare diagnosis in his list even in nonendemic areas.

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