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Case reports

Isolated primary orbital hydatid disease presenting as multiple cystic lesions: a rare cause of proptosis

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ABSTRACT. Echinococcal infection is a consequence of accidental ingestion of tapeworm eggs by humans. Liver scrutinises the initial haematogenous spread of portal blood and thus it is the most common organ involved. Isolated, primary involvement of other organs is a rarity. We describe a case of isolated orbital hydatid disease. To further add to the uniqueness of our case, two hydatid cysts were seen in our patient. The patient presented with unilateral proptosis with vision loss of the left eye since 6 months. Radiological imaging revealed two intraconal cystic lesions in the left eye. The chest radiography, abdominal sonogram and serology were negative for echinococcal disease. Surgical removal of the cysts was performed *via* lateral oribitotomy approach. Definitive diagnosis of *Echinococcus* was established on histopathology. The patient received post-operative albendazole therapy for 12 weeks. At 10 months' follow-up there was no recurrence or evidence of hydatid disease elsewhere.

Key words: *Echinoccocus granulosus*, hydatid, orbital, proptosis, radiology, cyst

Introduction

Hydatidosis is a parasitic infection caused by tapeworm *Echinococcus*. The echinococcal infection of human beings occurs *via* accidental ingestion of eggs passed in canine (definitive host) faeces which the cattle (intermediate host) ingest. Thus, the disease is endemic in areas where both dogs and sheep are reared together. Though the liver (60–70%) and lung (20%) are the most common sites for echinococcal disease, virtually any organ can be affected [1,2]. Uncommon sites of involvement include the heart, brain, muscle, salivary glands, spleen, pancreas, bone, adrenals, ovary and urinary tract [1]. Isolated, primary intra-orbital hydatid disease is extremely rare and represents around 1% of echinococcal cases [2–4]. Of these only 5% cases show multiple ipsilateral orbital hydatid cysts [5], as seen in our case. We thus highlight the importance of considering hydatid disease as a potential differential diagnosis of cystic lesion of the orbit. Moreover, we describe the imaging features and management for the same.

Case history

A 45 year-old-female presented with gradually progressive diminution of vision with proptosis of left eye since 6 months. There was no increase in size on bending forwards. Ophthalmic examination revealed non-pulsatile left sided proptosis, lid oedema, exposure keratitis and conjunctival injection (Fig. 1). The ocular movements were

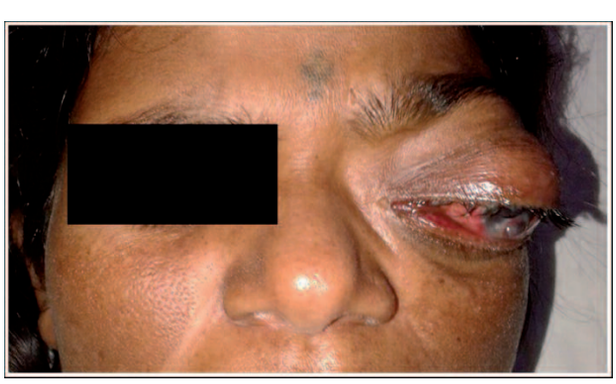

Fig. 1. 45-year-old female presenting with left sided proptosis, lid oedema, exposure keratitis and conjunctival injection of 6 months' duration

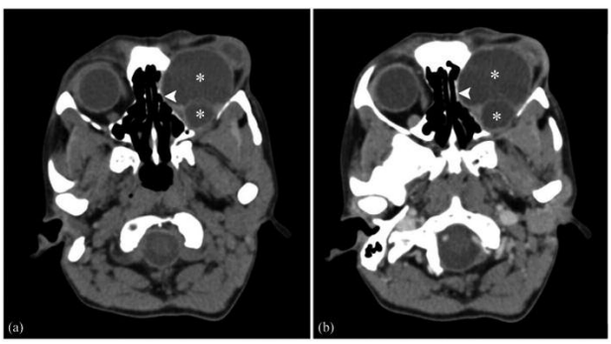

Fig. 2. Computed tomography, axial sections at the level of the orbit, unenhanced (a) and post-contrast images (b), show two well-defined, intraconal cystic lesions (*) within the left orbit with thinning of adjacent bones (white arrowheads)

restricted and painful. The visual acuity on the left side was 1/60. On fundoscopy, disc oedema was present. The right eye and remainder of the general examination was within normal limits. Routine blood and laboratory investigations were normal. Computed tomography of the orbit revealed two well defined, intraconal, fluid attenuation lesions placed one after the other on the left side of size 2.9×3.7×3.2 cm (Anteroposterior; AP×transverse; TR×craniocaudal; CC) and 1.2×1.1×1.4 cm (AP× TR×CC). The globe was pushed laterally and flattened from the posterior aspect by the cystic lesions. There was thinning of the adjacent orbital bones (Fig. 2). Magnetic resonance imaging showed T1 hypointense and T2 hyperintense lesions with mild post contrast rim enhancement (Fig. 3). Surgical excision was planned for our patient. A left-sided lateral orbitotomy was performed following which the cystic lesions were removed. Histopathological examination of the lesions was consistent with *Echinococcus granulosus* infection (Fig. 4). The patient had an uneventful post-operative period. The patient was discharged on post-operative day 10. She was put on oral albendazole (10 mg/kg/day for 12 weeks) and has been on regular follow up on an outpatient basis. Follow-up imaging at 6 and 10 months did not reveal any recurrence and currently the patient is symptom free. This case was considered as a primary isolated hydatid of the orbit as there was no previous history of hydatid disease and a complete radiological evaluation did not reveal hydatid infection at any other site. Chest radiography and ultrasonography of the abdomen performed after histopathological confirmation of echinococcal disease and at six months' follow up were negative for hydatid disease.

Discussion

Hydatid disease, caused by larval stage of *Echinococcus* tapeworms, is one of the most common zoonotic diseases worldwide and a major public health problem [1]. Isolated, primary orbital hydatidosis, without involvement of any other organ, as seen in our case, is very rare. Within the orbit, intraconal (as seen in our case) is the most frequent location. Besides, extraconal superior and inferior orbital region, subretinal space, extraocular muscles, lacrimal glands, vitreous and anterior chamber may also be involved [4]. Intra-orbital hydatid disease is generally unilateral with left side affected more than the right one [5]. Orbital hydatidosis is more common in children, with no sex predication [2]. Signs and symptoms include

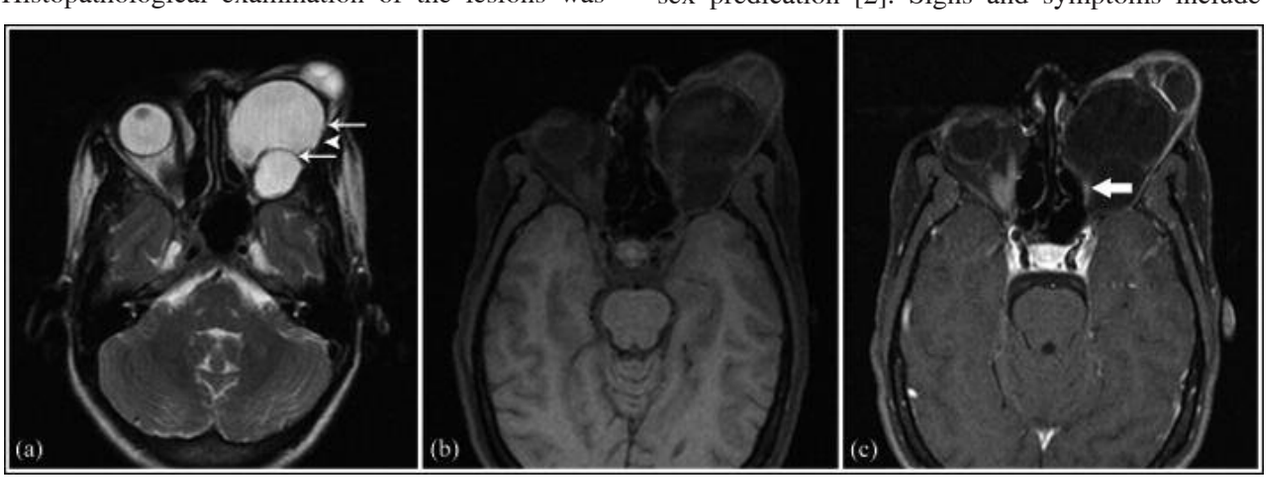

Fig. 3. Magnetic resonance of the orbit; shows two well-defined cystic lesions (thin white arrows) in the left orbit which are T2 hyperintense (a), hypointense on fat supressed T1 weighted images (b) and shows mild rim enhancement (thick white arrow) on post contrast T1 weighted images (c). Note the intraconal location of the cysts, medial to the lateral rectus muscle (white arrowhead).

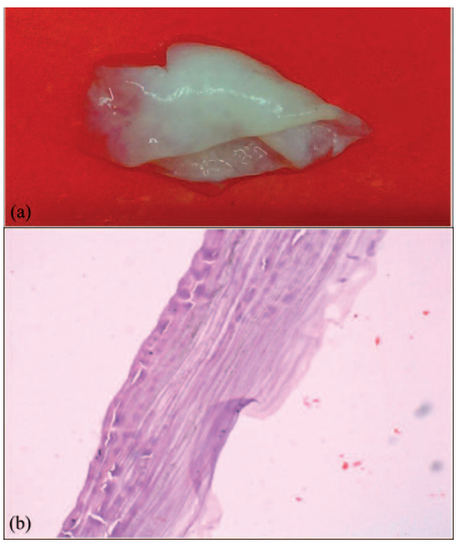

Fig. 4. (a) Gross pathological specimen showing the glistening translucent cyst wall. (b) Photomicrograph, hematoxylin and eosin stain (100×) shows the acellular laminated ectocyst of the hydatid cyst.

progressive proptosis with or without pain, visual impairment, periorbital pain, chemosis and headache. Other findings include restriction of ocular motility, diplopia, optic disc swelling, optic atrophy, palpebral mass and erosion of orbital bone. Orbital hydatid cysts are typically solitary lesions. Our case was unique in this matter as there were two cysts within the orbit. Multiple intra-orbital cysts have very rarely been reported in literature, a phenomenon which occurs in less than 5% of patients [5]. Radiological imaging shows a well-defined cystic lesion with absence of enhancing soft tissue. The double wall of a hydatid cyst may be appreciated on ultrasonography and fat suppressed sequences on MRI [4]. Presence of hydatid cysts in other organs and positive serological tests will further help. However, serological tests may be negative as far as isolated primary hydatid cysts are concerned. Definitive diagnosis is made only on pathological assessment [4]. Management is total surgical excision of the cyst [6]. Early diagnosis and treatment is promoted to prevent complications like spontaneous rupture and severe allergic response [2]. Surgery remains the primary treatment modality. Puncture-aspiration-injection-reaspiration (PAIR) technique has been described with little success [2]. Moreover, injection of hypertonic saline as a scolicidal agent protects against accidental rupture of the cyst [6]. Similarly, endonasal approach may be employed for cyst removal, although this has not been widely described in literature [3]. Medical therapy in the form of albendazole is added as an adjunct to surgery, and it is most effective if combined with praziquantel [4]. It is particularly useful if initiated 14 to 28 days before surgery [6]. Medical therapy is especially prescribed in cases of cyst rupture to prevent its possible dissemination and to prevent relapse [7]. Our case thus highlights multiple isolated primary orbital hydatid cysts in a 45-year-old female. We thus add to the differential diagnoses of cystic lesions of the orbit.

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