Case Report

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ORBITAL HYDATID CYST IN A PAEDIATRIC PATIENT - A RARE CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT

Hydatid cysts are commonly confined to the liver, lungs and rarely in the central nervous system, whereas localisation of hydatid cysts in the orbit are quite rare, accounting for less than 0.8 to 1% of all the cases. It is one of the rarest causes of proptosis even in endemic areas caused by dog tapeworm, Echinococcus granulosus. Definitive treatment is surgical excision, however complete removal is many times difficult. Present study discusses a case of a 12-year-old child who presented with a progressive proptosis of the left eye and was diagnosed with orbital hydatid cyst on histopathological examination.

KEYWORDS: Hydatid Cyst, Orbit, Echinococcus granulosus.

INTRODUCTION

Hydatid cyst, which us also known as human echinococcosis is a rare entity caused by the larval form of Echinococcus granulosus. Dogs are the definitive host whereas humans are the intermediate host where the larva develops (1-4). Human echinococcosis occurs due to the ingestion of food contaminated with eggs from which embryos are liberated into circulation (1-2). This tapeworm is common in hosts belonging to the parts of Middle East areas, Mediterranean, and regions of South America where it is endemic. (2) Most common organ involved is the liver (60-70%) and lung (10-15%), however, organs like the heart, kidney, spleen, thyroid gland, bones, and muscles are rare, amongst which orbital involvement is even rare and constitute less than one precent of all the cases (1-3).

CASE REPORT

A 12-year-old male child with complaints of progressive painless unilateral proptosis of the left eye for 1 year presented in the hospital. He also had mild restriction of movement of the left eyeball for past 6 months.However there was no history of trauma, diplopia, or diminished vision. He gave no history of close contact with cats and dogs. On local examination, the left eye was pushed downwards and laterally. Exophthalmometry revealed a difference of 7mm between the two eyes. Slight restriction in elevation of the eyeball and subconjunctival hemorrhage was noticed. Both sclera and the anterior segment were within normal limits and the pupils were similar bilaterally with normal reflexes. Also, the dilated fundus examination was within normal limits. Neurological examination was normal. Visual acuity of both eyes was normal with normal vision of 6/6.Other systemic examinations including the abdomen, lungs, and central nervous system were normal. Non-contrast Computed Tomography (NCCT) revealed a 32x19mm cystic mass which was well defined and located in the extra conal space on the medial and superior aspect of the orbit causing compression over the lamina papyracea of the left orbit. The orbit was displaced inferiorly and laterally and the mass effect was seen on the globe causing proptosis and a suspicion of Hydatid cyst was made. Routine blood investigations revealed eosinophilia. Abdominal ultrasonography, Chest X-ray was normal. The patient was taken for local excision of the cyst under general anesthesia. Excised specimen was sent for histopathological examination.On gross examination multiple yellow-colored membranous firm tissue pieces were received aggregate measuring 3x2cm (fig.1). Representative sections were taken and processed for histopathological examination that revealed lamellated eosinophilic membrane (fig.2) and focal germinal epithelium (fig. 3). No scolex or hooklets were identified. Based on clinical and histopathological examination a final diagnosis of Hydatid cyst of Orbit was made. Postoperatively the patient was treated with albendazole.

Fig. 1: Gross- Multiple yellow-colored membranous tissue pieces

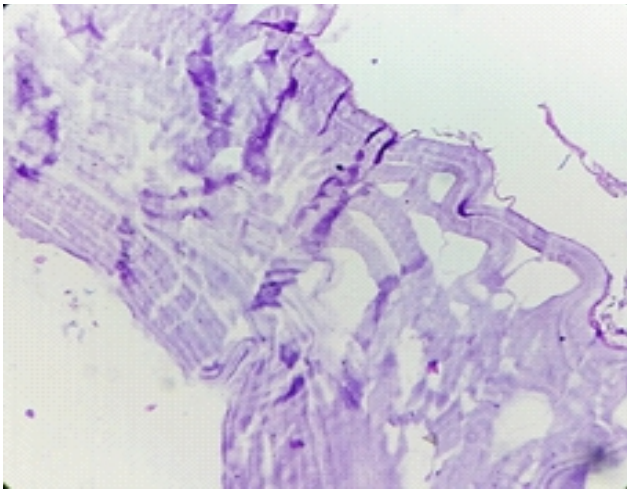

Fig. 2: Lamellated eosinophilic membranes with Germinal Epithelium (H&E-10X)

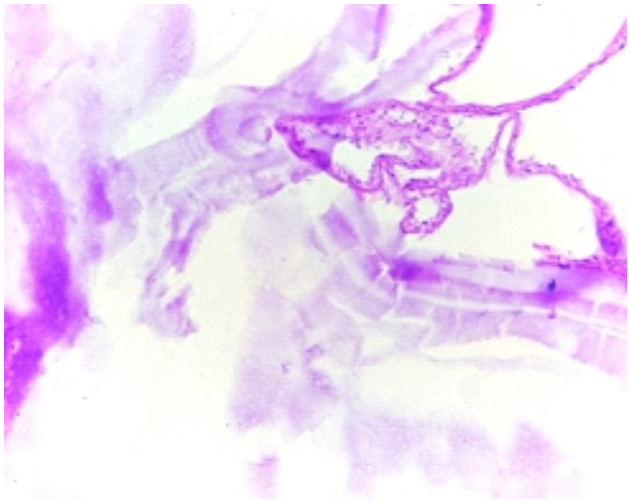

Fig. 3: Lamellated eosinophilic membrane with germinal epithelium lining (H&E- 40X)

DISCUSSION

Hydatid disease is caused by the metacestode which is the the larval stage Echinococcus granulosus (Dog tapeworm). (1-3) Although it is endemic in the regions of Middle East,South America, East Africa and parts of Australia, few cases are reported in non-endemic areas with international travel (1). No sex affliction has been reported (1). Orbital involvement is rare accounting for less than 0.8-1%, whereas intraocular hydatid cyst is extremely rare and may be confined to the retina, anterior chamber, and vitreous cavity (1). The disease results from the ingestion of contaminated food with the eggs of the tapeworm. The embryo is released in the intestine that attaches to the intestine, gains its way into the portal circulation, and settles down in tissues to form hydatid cysts in different organs (4). Cyst contains the scolices, hydatid fluid and brood capsules, in some cases (4).

Orbital cysts can be extraconal or intraconal in location. The superior quadrant is the most common extraconal location, whereas the intraconal location is very rare (1). The most common clinical findings are unilateral painless proptosis, restriction of ocular movements, and visual impairment (1, 4-5). Serological tests are usually negative. Eosinophilia occurs in 20-25% of cases (1, 4). On computed tomography, these cysts appear as unilocular hypodense, well-circumscribed, and thin-walled cysts. Fine peripheral enhancement and rim calcification can be seen following contrast (1-3). The differential diagnosis to be considered includes chronic hematic cysts, dermoid and epidermoid cysts, teratomas, and abscesses (1). Magnetic Resonance Imaging is definitive in which a hydatid cyst appears as low intensity on T1 and high intensity on T2 (1-3). .As an outcome of response of the body,a thick fibrous capsule surrounds the cyst which is known as pericyst. This Hydatid cyst wall is soft to firm, creamy white, shiny to glistening, and is made up of a laminated layer which is lined by a nucleated germinal layer, scolices and brood capsules develop form this lining (4-5). The diagnosis of hydatid cyst is mostly confirmed by histopathological examination of the cyst which shows three layers, an inner layer followed by the middle and the external layer. The middle layer which is the laminated layer is unique to the genus *Echinococcus* (6). In our case there was pearly white gelatinous cyst wall on gross examination and sheets of laminated eosinophilic layer on histopathological examination which confirmed our diagnosis. The only definitive treatment of hydatid cysts is complete surgical excision without rupture using Dowling's technique. (1-3) However, it as its own limitations due to the restricted area and complexity associated with orbital structures. (2,3) . Various other therapies like intracystic injections of hypertonic saline, 1% aqueous iodine, hydrogen

peroxide, 0.5% silver nitrate, and 10% formalin solutions can be used for destruction of the residual larvae (2-3). The most worrisome complication associated with the surgical removal of cyst is the rupture of the cyst, which may incite an anaphylactic reaction or may spread to the neighboring tissues(7). Drugs like Albendazole and Mebendazole have shown effective results in the treatment of hydatid cysts (8).

In conclusion, a Hydatid cyst must be considered a differential diagnosis, whenever a cystic lesion is encountered in the orbital region, especially in endemic regions. Timely diagnosis and complete removal of orbital hydatid cysts are very important to prevent any morbidity and mortality.

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