A Rare Case of Orbital Echinococcosis: A Histopathological Perspective

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ABSTRACT

Hydatid cyst of orbit is a rare disease constituting less than 1% of prevalence worldwide. It is a parasitic infestation by Echinococcus granulosus. It is prevalent in various regions of the world including South Asia. We, hereby, report a case of a 27-year-old male who was admitted to the ophthalmology ward of Jinnah Postgraduate Medical Centre. Presenting complaints were painless protrusion of left eye for the last few months progressing with time. After surgical excision, the specimen was sent to the Pathology Department, BMSI, JPMC, Karachi, for histopathological evaluation. Gross examination revealed multiple fragments of translucent, pearly-white, glistening thin wall of a cyst measuring 4x5 cm in aggregate. Histologically, features of hydatid cyst were identified with eosinophilic cyst wall, innermost germinal coat, containing attached and separated protoscolices and multiple daughter cysts, surrounded by dense fibrovascular tissue and chronic inflammatory infiltrate.

Key words: Orbital Echinococcosis, Orbital hydatid cyst, hydatid disease, HD histopathology

INTRODUCTION

Hydatid disease (HD) or Echinococcosis and also known as hydatidosis, is a cyclo-zoonotic parasitic disease caused by taeniid cestodes; a larval stage of Echinococcus.¹² The parasite typically maintains a dog-sheep-dog cycle and humans are accidently infected by ingesting eggs released from dogs.³ After ingestion, the larva gains entry into the blood circulation through intestines and seeding may occur in any part of the body, where it forms a cyst filled with fluid around itself.⁴ The disease is distributed worldwide and is considered a public health problem, however the disease is prevalent in regions notable for contact with cattle and sheep like central Asia, China, Australia, South America, Africa, and South Asia.⁵⁶ HD commonly affects liver in 60-70% of cases and lungs in approximately 20% of cases respectively. However Orbital involvement is rare, occurs in 1–2% of the cases, and usually affects children and young adults.⁷ Common clinical presentation of orbital Echinococcosis includes limited ocular movements, loss of vision, exudation, and oedema of eyelids and conjunctiva.⁸

The case with orbital Echinococcosis presented in outpatient department is reported below.
Case Presentation:

A 27-years-old male was admitted to the ophthalmology ward of Jinnah Postgraduate Medical Centre with painless protrusion of left eye for the last few months. According to him, the size of the swelling had progressed with time. He also complained of restricted eye movements with gradual loss of vision and exudative discharge from the same eye.

On ophthalmological examination, mild forward proptosis of left eye was observed with massive chemosis. Visual acuity was reduced in the affected eye, while it was normal in the right eye. Fundoscopy revealed papilledema of the left optic disc. The extraocular examination of the left eye revealed a single, large, well demarcated, lobulated, and cystic mass in the left upper palpaberal conjunctiva. The mass was non-tender and non-pulsatile. The bulbar conjunctiva revealed marked vascular congestion. The ocular movement was restricted on left side. Rest of the general and systemic examination was normal. However, serological and radiological imaging details were not provided by the ophthalmologists.

Surgical excision of the mass was performed and the specimen was sent to the Pathology Department of Basic Medical Sciences Institute, Jinnah Postgraduate Medical Centre, Karachi for histopathological evaluation.

The Specimen was fixed in 10% buffered formalin. The gross examination revealed multiple fragments of translucent, pearly-white, glistening thin wall of a cyst measuring 4x5 cm in aggregate. Histologically, the sections exhibited features of hydatid cyst revealing eosinophilic cyst wall, with innermost germinal coat, containing attached and separated protoscolices and multiple daughter cysts, surrounded by dense fibrovascular tissue and chronic inflammatory infiltrate (Figures A and B).

DISCUSSION

Hydatid disease or echinococcosis is a parasitic infestation by Echinococcus granulosus. Human is accidental host. The eggs are swallowed via water or vegetables contaminated with faeces of animals. They are then hatched within the small intestine and gain access to various organs through portal circulation. Unavailability of clean water and cattle breeding practices account mostly for this public health problem in Pakistan.9

To the best of our knowledge, we are presenting the first orbital hydatid cyst case in the past decade from the Pathology department, BMSI, JPMC. This disease is prevalent in younger population in endemic areas. In the present case, the patient’s age was 27 years. The mean age reported in literature was found to be 25.7 years. Patients are usually farmers or cattle breeders. However, no such history was provided in our case.10

Hydatid disease commonly affects liver followed by lungs while other body sites are rarely involved. Orbital hydatid cyst has been reported in less than 1% cases worldwide. The patient becomes symptomatic early in case of an orbital cyst, since there is less space for the cyst to expand. Other rare locations include intracranial, spinal, musculoskeletal, and cardiovascular systems.11

In the present case, no laboratory or radiological findings were provided. However, CT scan and MRI have proven to be the useful diagnostic tools in case of hydatid cyst. Histopathological examination provides a confirmatory diagnosis. Certain serological tests are
also performed for establishing a diagnosis as well as screening of this disease. These include enzyme-linked immunosorbent assay (ELISA), latex agglutination and indirect hemagglutination tests, replacing Casoni’s test. Furthermore, on imaging, the hydatid cyst may provide variable presentation and may pose challenges for radiologists. Thus, it is essential for radiologists from endemic areas to be able to identify the dynamic features of this cyst.

The treatment of choice for this lesion is surgical excision of the cyst. Utmost care is required to prevent the spillage of the cystic fluid into the surrounding tissue. In practice, the neighbouring area of the cyst is protected by 2% formalin. Cyst is punctured, fluid is aspirated, and then the walls are removed carefully.

In our case, the cyst was removed and fragmented segments of the cyst wall were submitted for histopathology. On gross examination, the cyst wall is thin, translucent, pearly-white, with glistening surface. Microscopic examination of the current case revealed the cyst wall composed of germinal layer. Protoscolices and a few daughter cysts were appreciated. Laminated membrane was also identified. Outermost layer was composed of fibrous tissue with scattered chronic inflammatory infiltrate. Similar features were reported by Nicholson et al. in their case series on orbital hydatid cysts.

Similar to a Turkish study, in our case, no significant postoperative complications were recorded.

CONCLUSION

Orbital hydatid cyst is a rare entity. It should be considered as a differential diagnosis in a cystic lesion or mass of orbit, where presenting symptoms include proptosis, chemosis, and blurred vision. Histopathologic examination is mandatory for confirmatory diagnosis. However, correlation with radiological imaging should also be taken into account.

Authors’ contributions: Nazish Jaffar conceived the idea and wrote the manuscript. Saba Sattar contributed in manuscript writing. Noshaba Rahat worked on histopathological diagnosis and details. Sadaf Razzaq carried out the literature search. Syed Mehmood Hasan critically reviewed the case. Saadia Akram edited and made the final review.

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