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CASE REPORT

Primary Multilocular Hydatid Cyst of Neck with Unique Presentation: A Rare Case Report and Literature Review

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Abstract Hydatid cyst (HC) is a parasitic infection caused by larva form of Echinococcus. It occurs frequently in liver and lungs. Primary HC of neck is extremely rare occurrence and here we report it with unusual presentation. A 35-year-old male presented with slowly growing painless swelling in right side of neck for 6–7 years. The swelling measured 11 × 6 cm and cough impulse elicited on local examination. MRI scan revealed a multiloculated cystic lesion in neck. Fine needle aspiration cytology yielded fluid aspirate and smear showed fragment of laminated membrane, suggestive of HC. The intact cyst was removed surgically and histopathologic examination confirmed the diagnosis of HC. Both clinicians and radiologists should consider HC in differential diagnosis of head and neck swelling for proper management.

Keywords Hydatid cyst · Neck swelling · Cough impulse · Multilocular cyst · Echinococcus

Introduction

Hydatid cyst (HC) is a zoonotic disease, caused by larva form of Echinococcus which may affect humans accidently [1]. HC develops most frequently in liver and lungs, rarely in other organs. Involvement of other organs can be primary or secondary following liver or lung involvement. Here we report a primary HC in neck of young male with unusual presentation, accidently diagnosed by fine needle aspiration cytology.

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Case Report

A 35-year-old male presented with slowly growing painless mass at right side of neck for the past 6–7 years. There was no history of fever, pain, dysphagia, odynophagia, hoarseness of voice or weight loss. Local examination revealed an oval shaped, 11 × 6 cm sized, soft, and non-tender mass. Cough impulse could be elicited on examination (Fig. 1). It did not move with deglutition or protrusion of tongue. The overlying skin showed no sign of inflammation. There was no other swelling found anywhere on the body. Physical examination including neurological and cardiovascular system revealed no abnormality.

Routine blood count, biochemistry and urine analysis were within normal limit. Chest X-ray and abdominal ultrasound were unremarkable. MRI scan of neck revealed a large multiloculated cystic lesion of 10.3 × 6.7 × 5.6 cm in size with septation and necrosis. Lesion was hypointense on T_1 weighted images and hyperintense on IR/T_2 images and was seen to extend inferiorly at the thoracic inlet indenting the lung cupola (Fig. 1). Fine needle aspiration cytology (FNAC) yielded 12 ml clear fluid with few white granular material which on microscopy showed fragments of laminated membrane and hooklets, suggestive of HC (Fig. 2). ELISA test for Echinococcus IgG antibody with patient's serum was found to be positive (1.05) which also favored our diagnosis (above 0.5 was positive).

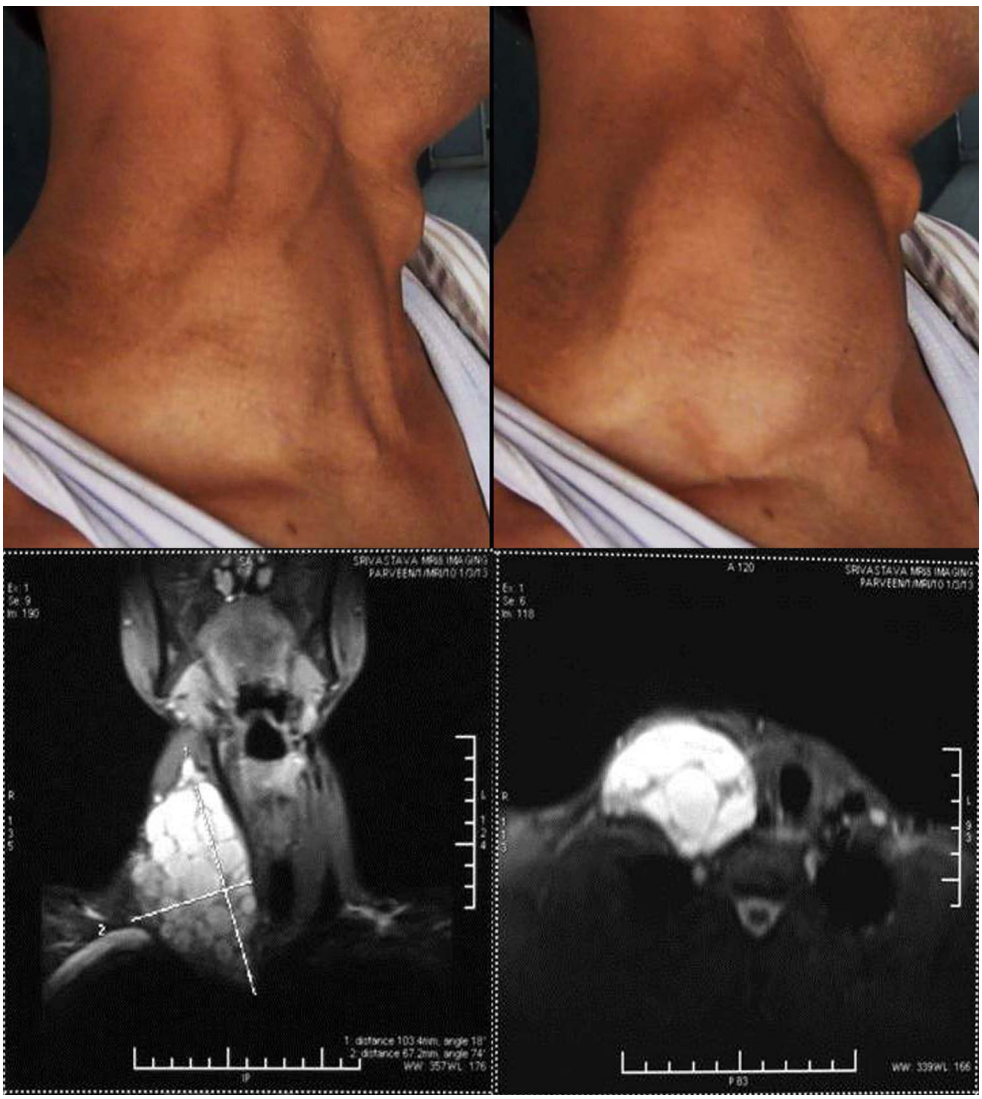

Fig. 1 Photographs showing preoperative view of neck swelling with cough impulse and multilocular cystic mass on T1 and T2 weighted MRI

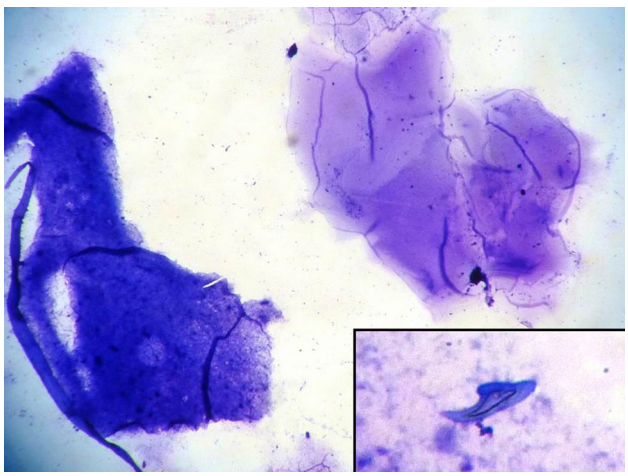

Fig. 2 FNA smear showing acellular laminated membrane of hydatid cyst along with hooklet in *inset* (MGG stain, ×10 & ×100)

Surgical excision of the cyst was then planned. Under general anesthesia the neck was explored and the cyst was seen extending inferiorly till the clavipectoral fascia and indenting upon the right lung cupola. Intact cyst was excised and sent for histopathological examination, which confirmed the final diagnosis of HC. Postoperative period was uneventful and the patient was kept on albendazole 800 mg/day for 4 weeks. The patient was followed up for 6 months and remained free of disease.

Pathological findings: Grossly, the cyst measured 9.5 × 6.5 × 4.5 cm, which on cut section showed multiloculation filled with white gelatinous membrane (Fig. 3). Hematoxylin and eosin stained sections showed numerous fragments of laminated, acellular ectocyst along with fibrous pericyst and inner germinal layer (Fig. 4). Final diagnosis of HC was made.

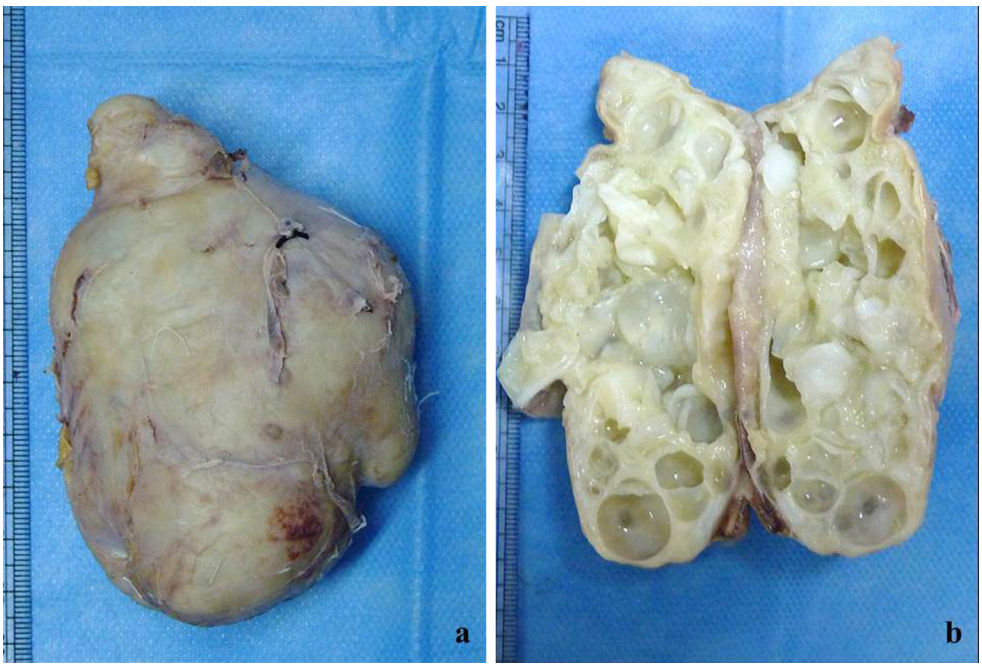

Fig. 3 Gross specimen of hydatid cyst showing multiloculated cyst on cut section

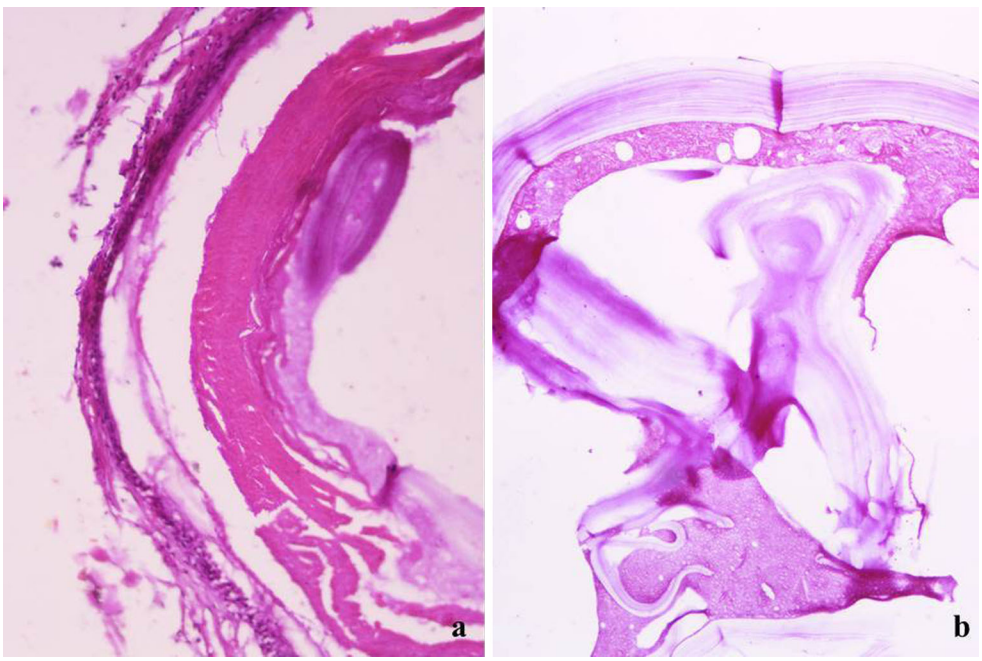

Fig. 4 Microphotograph showing pericyst and acellular laminated ectocyst of hydatid (H & E stain, ×10)

Discussion

Hydatid cyst, also known as hydatosis or echinococcosis, is a cyclo-zoonotic infection caused by the cestodes (tape worm) of genus *Echinococcus granulosus*, *E. multilocularis* and *E. oligarthus*. Of the two main forms of the HC, the unilocular cystic form caused by *E. granulosus* is far more common than the rare multilocular form caused by *E. multilocularis* [1]. The definitive hosts are dogs, wolves and foxes, while intermediate hosts are sheep, cattle, and horses [2]. Humans are accidental intermediate hosts and do not play any role in the biological cycle. For this reason, HC is endemic in cattle and sheep rearing regions of the world. Humans are occasionally infected by handling dogs

as well as by oral ingestion of Echinococcus eggs through contaminated food or water [2]. Majority cases come from rural areas or people who have settled in urban areas after spending life in rural areas as in our case [3]. In humans, infection is acquired by ingestion of eggs which hatch in the small intestine, pass into the portal venous system or lymphatic system to reach the liver and lungs and form HC. Moreover, they can cross the hepatic sinusoids or pulmonary capillary barrier to enter the systemic circulation and can affect any body parts [4]. Although the most commonly involved organ in human are liver (65–75 %) and lungs (15–25 %) but rarely 5–10 % cases can involve any organ of the body including heart, bone, muscles, spleen, kidney, brain, eye [2]. The occurrence of HC in head and neck region is extremely rare even in endemic areas [4, 5]. Multi-organ involvement is seen in 20–30 % of the cases [6]. So, patients having HC in any part of the body must undergo thorough systemic examination including liver and lungs. Our patient did not have any evidence of HC elsewhere in the body except the neck.

The majority of cases of HCs are asymptomatic although clinical signs and symptoms depend on the anatomic location, size and pressure effect of growing cysts. Therefore the signs and symptoms are variable and never pathognomic of HC [7]. So majority of HCs of neck are misdiagnosed on clinical examination. HC present as usually slow growing, fluctuant and painless mass [7]. As per our knowledge, we could not come across any report in English literature where cough impulse was associated with HC. The neck location in HC is itself is an unusual presentation. In our case, probably the cough impulse was transmitted impulse as the cyst was impinging inferiorly onto the apex of lung. Radiological investigations like ultrasound (USG), CT scan, MRI are useful diagnostic methods to visualize cystic masses by demonstrating daughter cysts, vesicle and internal septae [8]. But in our case, MRI could not reveal the precise diagnosis.

Fine needle aspiration cytology is an established diagnostic tool for evaluation of a neck mass. Although, it is not recommended in suspected case of HC due to possibility of acute anaphylactic reaction. However, some authors have reported that no complications were encountered during FNAC procedure [9]. We performed FNAC in our case as MRI finding was suggestive of multiloculated cystic lesion and exact etiology could not be ascertained. FNAC confirmed diagnosis of HC. Fortunately we did not encounter any complication. Different serological investigations for HC include ELISA, latex agglutination, direct hemagglutination, skin test (Casoni's test). Although serology is highly sensitive (80–100 %) and specific (88–96 %) for liver HC, it is less sensitive for lung (50–56 %) or other organ (25–56 %) involvement [9, 10]. However these tests are more valuable in the follow-up of treated patients.

On gross examination, HC is typically single and unilocular. However, multiloculated cyst can rarely be seen, as in our case. Histopathological examination is gold standard to make the final diagnosis of HC. The microscopy shows three layers of the cyst wall. The outer layer, known as pericyst, is a rigid protective layer which represents the host response to the parasite. The middle layer is white acellular laminated membrane. The inner germinal layer is thin and translucent [7]. Multilocular HC is composed of numerous small spaces or cavities, separated from each other by connective tissue. Each space filled with jelly-like matrix, mostly it is sterile but occasionally it may contain protoscolices. On extensive literature review, we found only few case reports on multilocular HC at various sites like kidney, myocardium, and uterus. The gross and histopathologic findings were similar as in our case.

Complete surgical removal of HC is the gold standard treatment [11]. The cyst should be excised as a whole without being ruptured to prevent recurrence or anaphylaxis [3]. Inactivation of daughter cysts and scolices before surgery can be achieved by injecting 20 % hypertonic saline, 5 % silver nitrate or formalin into cyst. Medical treatment alone is no effective but may be used if surgery is not performed because of multi-organ involvement and unapproachable location [9, 11]. Postoperative medical treatment with benzimidazole derivatives (albendazole, mebendazole) is frequently combined with surgery to prevent recurrence and high risk contamination [9].

Conclusion

We report this case for its unique presentation and unusual location. Imaging modalities, although sensitive, can sometimes not ascertain the exact diagnosis of HC. Thus, clinicians as well as radiologists should consider HC in differential diagnosis of neck swellings.

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