INTRODUCTION

Hydatid Cyst (HC) is a zoonotic disease caused by Echinococcus[1]. HC develops most frequently in liver and lungs, and rarely in other organs. Involvement of other organs can be primary or secondary to liver or lung involvement. Here we report a primary HC in submandibular region of a young female.

Case Report

A 34-year-old female presented with a slowly growing painless swelling at right side of jaw for past 10 years. There was no history of fever, pain, dysphasia, hoarseness of voice or weight loss. Local examination revealed a globular shaped, 8x6 cm, soft, and non-tender mass. Neither any signs of inflammation nor any cervical lymph nodes were found. Oral cavity examination was normal [Figure 1].

Blood investigations were within normal limit. Chest X-ray and abdominal ultrasound were unremarkable. Ultrasonography neck revealed a large cystic lesion of 8x6x5 cm size with septation. Contrast enhanced computed tomography scan of Neck showed a well-defined lobulated peripherally enhancing cystic lesion with internal enhancing membrane like structure in right side of neck suggesting possibility of hydatid cyst.

ELISA test for Echinococcus IgG antibody with patient’s serum was found to be positive. Surgical excision of the cyst along with the adjacent right submandibular gland was done. The cyst was seen extending to postero-medial stylo mastoid region involving posterior belly of right digastric muscle. Multiple thin membranes were found in the cyst cavity [Figure 2]. In postoperative period there were no signs of anaphylaxis with drain output of 80 ml serous on POD1 which gradually reduced and the patient was discharged on Albendazole 800 mg/day for 4 weeks. The patient was followed up for 4 months and remained free of disease.

Pathological findings

First specimen was of cyst in which Pericyst showed mixed inflammatory infiltrate comprised of lymphocytes admixed with eosinophils.

Second specimen was of right submandibular gland sections of which showed lobules of normal salivary glands.

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Vipul Kumar Srivastava, Shilpi Roy, Ram Nivas Meena and Rahul Khanna. Primary Hydatid Cyst of Submandibular Region

Figure 1 Clinical photograph of patient showing the swelling in right submandibular region caused by hydatid cyst.

Figure 2 Post-operative photograph of specimen demonstrating the germinal membrane of hydatid cyst excised from right submandibular region of neck.

DISCUSSION

HC, also known as hydatosis or echinococcosis occurs in 4 forms: cystic echinococcosis, caused by Echinococcus granulosus; alveolar echinococcosis, caused by E. multilocularis; polycystic echinococcosis, caused by E. vogeli and unicystic echinococcosis, caused by E. oligarthrus [1]. Of the two main forms of the HC, the unilocular cystic form caused by E. granulosus is far more common. The definitive hosts are dogs, wolves and foxes, while intermediate hosts are sheep, cattle, and horses. Humans are accidental intermediate hosts and do not play any role in the biological cycle. Humans are occasionally infected by handling dogs as well as by oral ingestion of Echinococcus eggs through contaminated food or water which hatch in the small intestine and pass into the portal venous system or lymphatic system to reach the liver and lungs[2].

Moreover, they can cross the hepatic sinusoids or pulmonary capillary barrier to enter the systemic circulation and can affect any body parts [3]. 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]. Multi-organ involvement is seen in 20–30 % of the cases [4]. Extra-hepatic HC although rare has been reported off and on in the literature. Ilica et al., 2007 have reported extra-hepatic HC to be present intraperitoneum, retroperitoneum, diaphragm, bone and soft tissue of the abdomen [5]. Most intraperitoneal cysts were secondary to spontaneous or iatrogenic rupture of hepatic, splenic or mesenteric HC. Primary peritoneal involvement was very rare. Primary splenic HC are also rare and most splenic cysts are secondary to systemic dissemination or intraperitoneal spread from ruptured hepatic HC. Similarly hydatid cysts of pancreas, ovaries, diaphragm and abdominal bone and soft tissue have been reported but most of them are secondary to hepatic or pulmonary HC.

Extra-abdominal HC is even rarer than extra-hepatic abdominal cysts. Curaya SY et al., 2012 reported primary HC of the gluteal muscle [6]. The first filter for the embryos from human intestines is the liver followed by the lungs. However embryos are known to pass through these two filters and primary HC although very rare can occur almost anywhere in the body.

Considering the rarity of a primary HC occurring in soft tissue of the neck, differential diagnosis of cystic lesions of the neck such as cold abscess, branchial cyst, plunging ranula, laryngocoele, lymphangioma, hemangioma, pseudoaneurysm, mucinous degeneration cysts, necrotic lymph nodes and last but not the least cystic metastatic lymph nodes should always be kept in mind.

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.

References
1. https://www.who.int/news-room/fact-sheets/detail/echinococcosis