CASE REPORT

Primary Hydatid Cyst of the Submandibular Gland: A Diagnostic Rarity

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Received on: 16 March 2022; *Accepted on:* 28 May 2023; *Published on:* xx xx xxxx

Abstract

Aim: To discuss the clinical presentation of a primary hydatid cyst of the submandibular gland and its surgical implications.

Background: Hydatid cyst disease is a parasitic, cyclo-zoonotic infestation by cestode of the genus *Echinococcus*. In humans, the most commonly affected organs are the liver (70%) and lungs (20%) followed by spleen, muscles, bones, kidneys, and the central nervous system. Primary hydatid cyst of the head and neck region; however, is extremely rare and very few cases have been reported in literature so far.

Case description: We report the case of a 28-year-old male who presented with a 3-year history of a gradually increasing swelling over the right submandibular region. It was firm, painless with no other relevant features. There was no history of exposure to farm animals or ingestion of tainted meat. Ultrasonography of the neck revealed a simple anechoic cyst followed by fine needle aspiration cytology (FNAC) which confirmed the diagnosis of a hydatid cyst. The patient underwent submandibular gland excision under general anesthesia wherein the entire gland with the cyst was excised in toto. He was also administered oral Albendazole (400 mg OD) over 4 weeks. The patient was completely asymptomatic at the 6 months and 1 year follow-up.

Conclusion: Though rare, hydatid cyst should be considered a differential diagnosis for submandibular gland lesions. Hydatid cyst fluid (HCF) is highly antigenic and may cause allergic reactions ranging from mild hypersensitivity to severe anaphylaxis which may be potentially fatal for the patient. This may occur spontaneously, due to accidental trauma or iatrogenic manipulation. Hence, great precision and precaution need to be exercised during the surgical excision to prevent any spillage of contents in the surgical field.

Clinical significance: Being extremely rare in the head and neck region, hydatid cyst is liable to be misdiagnosed unless there is a high degree of suspicion. It is imperative that a hydatid cyst be preoperatively diagnosed and positively confirmed as any accidental or iatrogenic trauma may cause release of the HCF. Also, the intraoperative dissection needs to be meticulous and precise and arrangements should be made in anticipation of complications to ensure a favorable outcome for the patient.

Keywords: Case report, Fine needle aspiration cytology, Head and neck region, Hydatid cyst, Imaging, Primary, Submandibular gland, Surgery, Ultrasonography.

Otorhinolaryngology Clinics: An International Journal (2023): 10.5005/jp-journals-10003-1467

Introduction

Hydatid cyst disease is a parasitic, cyclo-zoonotic infestation by cestode of the genus *Echinococcus*. It commonly presents as either cystic or alveolar echinococcosis. The three most commonly encountered species are *E. granulosus, E. multilocularis*, *and E. vogeli*. Dogs and other canine species[1] are the primary or definitive host for the parasite whereas other mammals, such as sheep, cattle, horses, and occasionally humans act as the intermediate hosts for this taeniid parasite. Globally, the disease is rare, but it is endemic in regions where cattle rearing or sheep farming is practiced, including the Mediterranean countries, Middle East, parts of Central Asia including many regions of China, India, southern regions of South America, southern and central regions of Russia, and parts of South America and Africa. Even in endemic regions, however, primary cystic echinococcosis of head and neck region is very rare.[1–4] Cases have been reported of echinococcal infestation of the maxillary sinus, parapharyngeal space, infratemporal fossa, pterygopalatine fossa and salivary glands. Among these, hydatid disease of the cervicofacial region, especially salivary glands is one of the most uncommon entities encountered. We discovered only 14 reported cases of primary hydatid cyst of the submandibular gland in literature (Table 1).[1,5–17]

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How to cite this article: Ekhar VR, Nayak P, Shelkar R. Primary Hydatid Cyst of the Submandibular Gland: A Diagnostic Rarity. Int J Otorhinolaryngol Clin 2023;x(x):xx–xx.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

In view of its rarity, we present here a case of primary hydatid cyst of the submandibular gland. It begs to remind us that hydatid cyst should be considered a differential diagnosis for cystic

Table 1: Reported cases of primary hydatid cyst

	Patient demographic	*Treatment instituted*
Singh S (1972)[5]	50 Y/M	Submandibular gland excision
Onerci et al. (1991)[1]	41 Y/F	Submandibular gland excision
Khademi et al. (1996)[6]	46 Y/F	Submandibular gland excision
Kini et al. (1997)[7]	14 Y/M	Excision of the lesion
Bouckaert et al. (2000)[8]	20 Y/F	Excision of the submandibular gland and surrounding lymph nodes
Sahni et al. (2000)[9]	28 Y/F	Submandibular gland excision
Geopgopoulos et al. (2007)[10]	56 Y/M	Submandibular gland excision
Pal and Shankar (2008)[11]	52 Y/M	Submandibular gland excision
Daneshbod and Khademi (2009)[12]	46 Y/F	Submandibular gland excision
Bhatia et al. (2010)[13]	7 Y/M	Submandibular gland excision
Karmarkar et al. (2011)[14]	30 Y/F	Excision of the submandibular gland
Berkiten et al. (2013)[15]	45 Y/F	Excision of the submandibular gland with marginal mandibulectomy and excision of the fistulous tract
Manandhar et al. (2015)[16]	16 Y/F	Submandibular gland excision
Chickhladze (1959)[17]	6 Y/F	Submaxillary gland excision

swellings in the head and neck region even without systemic involvement.

Case Description

A 28-year-old male presented to the outpatient department of ENT, in a tertiary care setup with a 3-year history of a gradually enlarging swelling in the right submandibular region. Examination revealed a firm, painless, bimanually palpable mass of about 4 × 3 cm in the right submandibular area. There were no systemic complaints, no history of swelling over any other region, cough, or hemoptysis.

Suspecting a submandibular gland lesion, we proceeded with ultrasonography of the swelling. It showed evidence of a simple, anechoic, cystic lesion within the submandibular gland of size 3.6 × 2.5 cm, with no connection to the submandibular duct suggestive of a benign cyst. Based on these findings, fine needle aspiration cytology (FNAC) was performed. The aspirated fluid showing fragments of the parasite consisting of scolices, hooklets in a background of proteinaceous material and laminated membrane suggestive of a hydatid cyst. On further enquiry, the patient gave no definitive history of exposure to farm animals or ingestion of tainted meat. There was no history of similar complaints in the family either.

The complete blood count was normal with no evidence of eosinophilia. Erythrocyte sedimentation rate and other routine hematological investigations were also within normal limits. Chest X-ray and abdominal sonography revealed no relevant abnormal features suggesting any other lesion, leading to the diagnosis of a primary submandibular hydatid cyst.

The patient was taken up for surgical excision under general anesthesia after a short course of Albendazole 400 mg OD for 5 days. Intraoperatively, the mass was seen to occupy almost the entire submandibular gland necessitating the removal of the gland (Fig. 1) with the cyst in toto without any rupture or spillage of the fluid. The specimen was sent for histopathological examination. On gross, the section showed the various layers of the cyst including the laminated ectocyst wall and the daughter cysts contained within (Fig. 2). Stained sections too correlated with and confirmed the diagnosis of a hydatid cyst by *E. granulosus* (Figs 3 and 4).

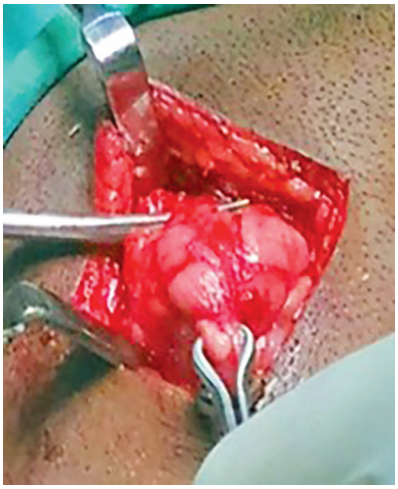

Fig. 1: Intraop photo showing a multiloculated cyst in the submandibular salivary gland

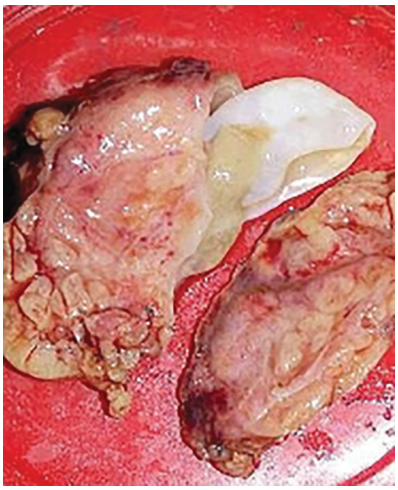

Fig. 2: Gross specimen showing the laminated membrane of the cyst

Fig. 3: Stained histopathological section of the hydatid cyst showing the lamellated ectocyst

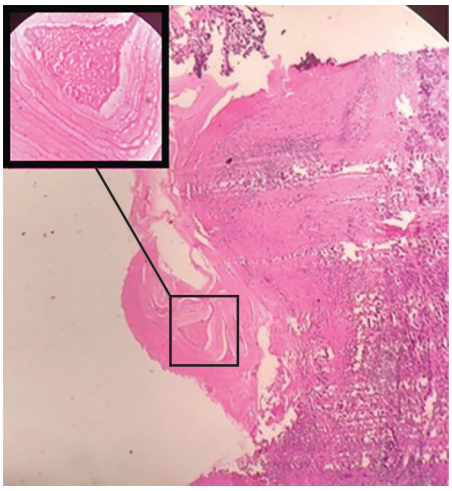

Fig. 4: Magnified view showing multiple daughter cysts contained within the lamellated membrane [inset]

Postoperatively, the patient was prescribed albendazole (400 mg OD) for 4 weeks.

The patient was discharged on the third postoperative day was completely asymptomatic at 6 months and 1 year follow-up. Follow-up ultrasonography did not reveal any lesions in any other organs.

Discussion

Primary hydatid cyst of head and neck is an exceptionally unusual entity accounting for only about 2% of all cases,[2,4] even in endemic regions. It poses a dilemma in terms of diagnosis due to the rarity of its anatomical location. It may cause substantial mortality and morbidity if misdiagnosed or improperly managed. The adult parasite resides in the bowels of the definitive host whereas the intermediate host harbors its larval form. The intermediate hosts, in this case, humans, acquire the disease by ingestion of the eggs of the parasite through contaminated food. The eggs then hatch, penetrate the intestinal wall, reach the liver and then disseminate to various organs. In humans, the most commonly affected organs are the liver (70%) and lungs (20%)[4] followed by spleen, muscles, bones, kidneys, and the central nervous system.

Hydatid cyst is essentially diagnosed on the basis of a detailed clinical history (including contact with animals), a thorough physical examination aided by radiological modalities, cytological evaluation and serological tests. Clinically, hydatid cysts are slow growing, painless lesions with no pathognomonic signs and symptoms, especially in the head and neck region. Clinical manifestations if present are due to either pressure effect of the cyst in the surrounding tissues or allergic reaction to the fluid within if ruptured. As involvement of the head and neck region is uncommon, a comprehensive systemic examination must be executed to rule out involvement of more common organs like liver and lungs. Multiorgan involvement besides liver and lungs must also be ruled out as the prevalence of the same has been found to be as high as 20–30%.[4,18] In our case, however, the patient had no systemic signs or symptoms and no clinical manifestations besides a slow growing swelling in the right submandibular region thus cinching the diagnosis of a primary submandibular hydatid cyst.

Radiological techniques like USG, CT, and MRI play a very important role in preoperative diagnosis of hydatid cyst. They may identify the multiple loculi, thin-walled membranes, internal septations and the contained daughter cysts. CT may depict the characteristic spoke-wheel appearance, the "water-lily" sign. Old, inactive cysts may show calcification.[4] CT has one of the highest sensitivity and specificity as a diagnostic modality for identification of a hydatid cyst. In this case, CT was not done as the ultrasound and clinical examination gave no hint of any differential diagnosis but a simple cyst.

Serological investigations like agglutination test, ELISA and immunoelectrophoresis have low sensitivity and specificity but may provide a useful substitute for the more expensive radiological investigations. They may be used to recognize carriers, assess response to treatment and monitor prevalence of the disease.[19,20] These investigations were not done in our patient as the patient was penurious and these tests are unavailable in our setup.

Fine needle aspiration cytology though controversial, as it may precipitate anaphylactic reactions[2] and cause spread of daughter cysts, has been used by many doctors to diagnose hydatid cyst in recent times without any ill-effects.[21] Fine needle aspiration cytology was performed on our patient using a 25-gauge needle, to clinch the diagnosis as the initial USG performed was inconclusive. The patient suffered from no ill-effects due to the same.

Surgical excision remains the most effective method of management of a hydatid cyst[22] A preoperative course of anti-parasitic agents like albendazole and praziquantel may reduce the size of the cyst and render the cyst sterile and thus reduce complications during surgery. Arif et al. observed that a 12-week course of albendazole preoperatively significantly reduced the viability of the cysts during surgery and also reduced the recurrence rate considerably.[23] Albendazole has also been used postoperatively in patients after excision in many cases.

PAIR or "puncture, aspiration, injection and re-aspiration" technique has been used successfully for treatment of hydatid cyst in individuals with liver, bone or muscle cysts,[24] unfit for either surgery or anesthesia. It is performed under ultrasound or CT guidance, involving aspiration of the cyst contents via a cannula, followed by injection of a scolicidal agent, and then re-aspiration of

the contents. This is repeated until the return is clear. There are also reports in literature of it having been used in parotid, submandibular and thyroid hydatid cysts without any intraoperative spillage and a good long-term prognosis. To overcome minor drawbacks like difficulty in aspiration of solid contents of the cyst, a variation of PAIR called percutaneous techniques (PT) is being more widely utilized in recent times. Surgical treatment however is the only definitive treatment of a hydatid cyst.[25]

Clinical Significance

Being extremely rare in the head and neck region, hydatid cyst is liable to be misdiagnosed unless there is a high degree of suspicion. Clinically, it is imperative that a hydatid cyst be preoperatively diagnosed and positively confirmed as any accidental or iatrogenic trauma may cause the release of the hydatid cyst fluid (HCF). Hydatid cyst fluid is highly antigenic and may cause allergic reactions ranging from mild hypersensitivity to severe anaphylaxis which may be potentially fatal for the patient. Once diagnosed, the patient should be made aware of the complications, especially the symptoms related to anaphylaxis. They should be counseled to watch out for symptoms like pruritis, urticaria, flushing, edema, dyspnea, cyanosis, and light-headedness.[26]

The incidence of anaphylaxis during the surgery for hydatid cyst excision is very low. Studies report it to be between 0.2 to 3.3%.[27] However, mortalities have been reported sporadically during surgical excision. The intraoperative dissection therefore needs to be meticulous and precise. Arrangements should be made in anticipation of complications. A close monitoring should be done for persistent desaturation, hypotension, hypocarbia, bradycardia, and arrythmias during the procedure due detect hemodynamic collapse or increase in airway pressure. In case of any spillage in the operative field or release of HCF into the systemic circulation with subsequent symptoms of shock, immediate administration of intravenous (200 ug) or intramuscular (0.5 mg) adrenaline, intravenous corticosteroid and chlorpheniramine maleate (45.5 mg) is recommended. Besides these, the intravascular volume, vessel tone, and cardiac output should be supported by fluids (crystalloid or colloid). Furthermore, inhalational anesthetic agents should be withheld, and 100% oxygen ventilation should be commenced to counter airway compromise due to bronchospasm, if present. Extubation should be deferred until the airway integrity is confirmed. Prophylactic steroids and antihistamines may be administered to prevent exacerbation or recurrence of the symptoms.[28]

In conclusion, hydatid cyst, though rare, must be considered a differential diagnosis for cystic lesions of head and neck, especially in endemic regions to ensure appropriate management without any morbidity to the patient. Early diagnosis and treatment are essential to ensure a favorable patient outcome.

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Ethical Approval

Ethical approval was waived by the Institutional Ethics Committee of Indira Gandhi Government Medical College in view of the retrospective narration of the case study and all the procedures performed were part of the routine care.

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