Radiological Diagnosis of Sub Mandibular Region Hydatid Disease: A Case Report and Literature Review

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Abstract

The rarity of primary hydatidosis in sub mandibular neck region may create a diagnostic dilemma with brachial cyst, dermoid cyst, sebaceous cyst and even lipoma. Even in endemic population with frequently reported hydatid disease, the diagnosis of submandibular region hydatidosis needs a high index of radiological suspicion like double capsule pattern on ultrasonography and multiloculated hypoechoic cystic disease with scolices seen on computed tomography scans. An experienced radiologist may report these rare diagnoses and avoid aspiration cytology thus decreasing chances of theoretical anaphylactic reactions. Different clinical presentation of primary hydatidosis depends on the cyst load, topographic location of cysts and the host’s immune response. We report a case of an 18-year-old boy with soft, painless left submandibular region swelling since eight years, who was diagnosed radiologically to be a case of hydatid disease of submandibular gland region and successfully underwent surgical excision without preoperative fine needle aspiration cytology. The review of literature suggest no definite consensus regarding routine use of aspiration cytology for diagnosis but we here by propose that proper preoperative radiology may avoid need of any cytology whatsoever.

ABBREVIATIONS

USG: Ultrasonography; CT: Computed Tomography; FNAC: Fine Needle Aspiration Cytology, ELISA: Enzyme Linked Immune Sorbent Assay; CXR: Chest Roentgenogram

INTRODUCTION

Hydatid cyst is a zoonotic parasitic disease caused by Echinococcus Granulosus where dogs and wolves are the definitive host and the intermediate hosts are sheep and goats while humans are accidental intermediate host. The cattle and sheep rearing areas of South America, Middle East countries, India, southern and central Russia, and many parts of China are the endemic population for hydatid disease of human beings [1,2]. Primary hydatid disease of head and neck region is a rare occurrence even in these endemic areas. Liver and lung comprise two of the most common location but once larvae crosses the hepatic sinuses and pulmonary capillaries barrier, it can dislodge anywhere, leading to systemic dissemination. In Indian subcontinent owing to its poor hygiene, close interaction with domestic animals, overcrowding and frequent consumption of poorly cooked meat, further contributes to the disease burden.

CASE PRESENTATION

An 18 Year old muslim male (weight-50 kg, height-168 cm) from poor socio economic background, residing in crowded locality of western uttar-pradesh province in India, presented to outpatient department with a swelling in the left submandibular region since last eight years, which was insidious in onset, gradually progressive and painless. There was no history of any trauma or previous surgery. There was no history of fever, weight loss, night sweats or any contact with any patient of tuberculosis. Personal history revealed proximity with pets and cattles at home. On clinical examination, there was a uniform 4 x 4 centimeter, well defined, round, globular, soft, non-tender swelling over left submandibular region. It was neither adherent to the underlying structure nor fluctuant. The overlying skin was smooth, normal looking, pinchable and without any scars or sinus. There was no other swelling in the neck. Examination of oral cavity, ear, nasal cavity, pharynx revealed no abnormality. Systemic examination was normal. Preliminary blood tests showed normal counts and eosinophilic sedimentation rate. With a differential diagnosis of dermoid/brachial cyst/cold abscess/sebaceous cyst an Ultrasonography (USG) was planned that showed normal counts and eosinophilic sedimentation rate. With a differential diagnosis of dermoid/brachial cyst/cold abscess/sebaceous cyst an Ultrasonography (USG) was planned that revealed thick walled, isoechoic, cystic mass with septations and classical double capsule pattern pointing towards hydatidosis (Figures 1,2). Thus a radiological surprise was confirmed by computed tomography (CT) scans. The CT scan showed well...
defined hypodense, 3x3 centimeter, round, encapsulated, multicystic lesion abutting posterior part of left submandibular gland without any compression of vital structures (Figure 3, Link 1). A provisional diagnosis of hydatid cyst was made and fine needle aspiration cytology (FNAC) was avoided, as there exists a theoretical risk of spread of hydatid disease and anaphylaxis. To rule out liver or other systemic involvement a USG of abdomen and chest roentgenogram (CXR) was done which was normal. Serology test was differ in view of its poor sensitivity and specificity in extrahepatic hydatidosis and poor socioeconomic status of our patient. With a provisional diagnosis of primary submandibular gland region hydatidosis, excision was planned under general anesthesia using a left transverse cervical incision two finger breadth below body of mandible. The muscles and fascia were dissected and cyst was exposed. It was aspirated and injected with hypertonic saline and reaspirated. The cyst was adherent to poster inferior part of left submandibular gland and while dissection there was inadvertent cyst rupture and spillage. Pale whitish, cheesy material was seen which was collected and sent to pathology along with the ruptured cyst. Antibiotic mixed hypertonic saline (20%) irrigation of cavity was done thoroughly. The incision was closed in 2 layers over a suction drain. Histopathological examination report confirmed the diagnosis of hydatid cyst by demonstrating a germinal layer with a lamellated ectocyst with fibrous outer layer. Also significantly noted was marked foreign body type of giant cell reaction around the pericystic layer that was reaching the adjacent fibromuscular tissue, thus confirming hydatid disease. The patient was given one month course of tablet albendazole (400mg) twice daily and was symptom free at 3 months follow up for both the local site and the usual primary sites of liver and lung as seen on follow-up CXR & USG abdomen.

**DISCUSSION**

Echinococcosis, being a zoonosis, occurs primarily in sheep-grazing areas of the world, but is common worldwide because dog is a definitive host. Humans contract the disease from dogs, but there is no human-to-human transmission [2]. The adult parasite (0.5-1 cm) can reside in intestines of dogs, wolves, foxes and jackals. Ova being resistant to various environmental conditions are excreted in their feces and either herbivores or humans ingest the infected food materials. These ova after hatching in small intestine, penetrates the mucosal walls, reaching the liver (66%) via portal vein. The larvae that pass through this first filter, reaches lung (5-15%) via right heart [3]. Subsequent passages through this second filter now cause systemic hydatidosis [4,5]. Various case series have cited frequency of extra hepatic and extra pulmonary hydatidosis to be approximately 9%. Even in endemic areas the musculoskeletal hydatidosis has only been reported to a tune of 0.5% to 3% and almost always secondary to hepatic or pulmonary dissemination [7]. The largest ever reported case series on hydatid disease has only reported 2.3% (24 out of 1056) cases of soft tissue involvement. Literature review shows various genetic and phenotypic variations that have been demonstrated in *E. granulosus*, and molecular tools have identified almost 10 different genotypes (G1–G10). *Echinococcus granulosus* sensu lato is considered to be a species complex, comprising of *E. granulosus* sensu stricto (G1–G3), *E. equinus* (G4), *E. ortleppi* (G5), *E. canadensis* (G6–G10) the 'lion
Our case created interest as it was a primary hydatid cyst involving submandibular gland region without liver or lung involvement or ruled out on radiology and very few similar cases have been reported previously worldwide [10,11]. Its very difficult explaining, how the larva escaped two filters and formed a solitary neck swelling, hence a possibility of lymphatic spread can be kept at times. The initial presentation hardly points towards hydatidosis even in endemic areas and thus detailed history including occupation, residence and family history becomes essential. Primary clinical examinations usually points toward differential diagnosis of cold abscess and dermoid and other more common pathologies like branchial cyst and there are no pathognomonic pointers to hydatid disease. For serology, literature review revealed two schools of thought where few advocates FNAC as gold standard for diagnosis while others reported FNAC to cause infection of the cyst, dissemination and anaphylaxis even with aseptic precaution [12-14]. It is here when the role of radiological diagnosis surpasses serological diagnosis. The non-invasive nature of tests and pathognomonic pointers like double capsule pattern in a cystic, a vascular lesion having internal septations with the presence of daughter cysts on USG and hypodense, thick walled, well-delineated cystic lesion having internal septations, with or without bony erosion on CT scan clinches the diagnosis [14,15]. The CT-scans have an accuracy of 98% in demonstrating the daughter cysts.

Although various serological tests are performed in abdominal hydatidosis like latex agglutination, skin test (Casoni intradermal test), hemagglutination and ELISA, they have low diagnostic sensitivity (only 50%) and specificity in extra hepatic disease, as in head and neck hydatidosis [16]. Eosinophil count is also seen to be high in only 25% of the cases, while hypogammaglobulinemia is noted in only 30% cases. These are mainly used in follow-up to detect recurrence if increasing titres are noted.

Surgery remains the main stay of treatment and in toto excision without spillage of content is the key. In cases with preoperative definitive diagnosis, the techniques of preliminary aspiration and instillation of a combination of hypertonic saline (20%), formalin and silver nitrate (0.5%) has been used to inactivate the protoscoleces and prevent seedling. Since there was intraoperative spillage of cyst, we preferred thorough antibiotic mixed saline irrigation of the cavity. Although the Patient was discharged on oral Albendazole tablets (400 milligrams) twice a day for one month, there are variable protocols for medical management and follow-up after surgery. There are centers treating patient with Tab. Albendazole (400mg BD) before surgery for 21 days and post-operative same dosing schedule for 2 to 6 months. Pre-operative albendazole, based on aspiration cytology diagnosis has been proposed to prevent recurrences [17]. The chemotherapy makes the daughter cysts inactive and dead and is mainstay of treatment in cases with unapproachable surgical sites and multiorgan involvement. At three months of follow up, our patient showed no sign of local as well as primary site recurrence as evaluated on CXR & USG abdomen.

To conclude, Primary hydatidosis can occur anywhere from head to toe [18]. Hydatid disease can be a differential diagnosis of cervical mass especially in endemic countries. Fine needle aspiration cytology is better avoided if high suspicion of echinococcosis is there, as risk of anaphylaxis is a major concern. Also there is aned to rely on radiological diagnosis, as any invasive procedure may complicate the complete treatment procedure.

Great care must be taken to avoid spillage of cyst content during surgery and appropriate precaution should be taken in case the cavity is contaminated by any inadvertent injury. Histopathological examination and patient follow-up is critical in all cases for an accurate diagnosis, definitive treatment and to prevent recurrence.

REFERENCES


