Case Report

Hydatid cyst of the buccal mucosa: An unusual presentation

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Abstract

Hydatid cyst is a parasitic cyst caused by the tapeworm Echinococcus that occurs primarily in sheep grazing areas worldwide. It is a chronic disease, and the cysts can be localized in unusual anatomical and geographic locations. It is known to affect the head and neck region. Patients must undergo a thorough systemic investigation as 20–30% show multiorgan involvement. We report a case of hydatid cyst occurring in the buccal mucosa of a 45-year-old male presenting as a small asymptomatic lump and emphasize on its rarity and diagnostic issues.

Key words: Buccal mucosa, echinococcosis, hydatid cyst

INTRODUCTION

Hydatid disease or cyst is a parasitic infection caused by the cestodes of genus, hence also known as Echinococcosis. Echinococcus E.chinococcosis granulosus is the most common form that causes unilocular hydatid cyst.[1] Hydatid in Greek means “drop of water”. [2] Dogs are the definitive host. Cattle, pig, sheep, and occasionally humans are the intermediate hosts. Hence, sheep and cattle raising regions have the highest incidence of this disease, especially in Africa, New Zealand, South America, Central Europe, and the Middle East Australia.[3]

The infestation is by direct contact with dogs commonly and ingestion by food contaminated with dog feces. It is said to be acquired usually in childhood.[2] Hydatid cysts affect humans, and the most frequent localizations are the liver and lungs (80%). They have also been reported to occur in the peritoneum (20%), spleen (0.7–8%), kidney (7%), skin and muscles (4%), nervous system (0.2–3%), bones (2%), and heart (0.2–2%).[1] It is estimated that about 1–2% manifest in the head and neck region, involving the parotid and submandibular glands, tongue, maxillary sinus, infratemporal fossa, parapharyngeal space and pterygopalatine fossa as well as the posterior or anterolateral cervical regions have been reported in the literature.[2,4] This rare occurrence is even in countries where Echinococcus infestation is endemic. In the past, this was an unusual disease but now it is no more considered to be restricted to a defined population in localized geographic regions and is reported to occur anywhere.[5] The need to be aware of its clinical presentation, diagnosis, and management is all more acute as the clinical features vary according to the location and size of the cysts. Only two cases of hydatid cyst involving buccal mucosa having been reported in the literature. Third case of an asymptomatic hydatid cyst localized in the buccal mucosa of the oral cavity in a 45-year-old male is reported here.

CASE REPORT

A 45-year-old male reported to the dental college for a regular dental checkup. An accidental finding of a reddish brown swelling was revealed on clinical examination over the buccal mucosa. On inspection, it was a well-defined solitary oval shaped swelling with bluish translucency and with no signs of inflammation, discharge or bleeding. The patient was aware of
the lesion but considered it insignificant due to its asymptomatic nature. The swelling measured about 1 cm × 1 cm extending from 34 to 35. It was sessile, soft in consistency, fluctuant, and partially tender on palpation. A provisional diagnosis of mucocele was given [Figure 1]. Excisional biopsy was done and was sent for histopathological evaluation. Gross morphology of the specimen showed a whitish punctum [Figure 2]. The hematoxylin and eosin stained sections revealed a cyst which was trilaminar made up of the outermost adventitial layer/pericyst, the innermost germinal layer/endocyst, and the intermediate avascular eosinophilic refractile chitinous layer/exocyst [Figure 3]. The lumen was basophilic (necrotic) showed numerous vesicular projections on its germinal layer [Figure 4]. The vesicles or the brood capsules contained scolices in various stages of maturation and hydatid sand demonstrating hooklets were seen [Figure 5]. However, we could not identify any definitive scolices, and this made us difficult to identify the species. The cyst ruptured after the removal of the luminal contents. A histopathological diagnosis of hydatid cyst of the buccal mucosa was given. Routine hematological examinations revealed no abnormalities. The patient was then started on albendazole at 15 mg/kg/day for a month. A chest radiograph and abdominal sonography scans were requested. No hydatid cysts were identified anywhere else in the body. The patient is on follow-up till date with no recurrences.

**DISCUSSION**

Hydatid cysts are characteristically slow growing and asymptomatic benign cystic lesions affecting both men and women in equal proportions. It is seen that the interval between the initial infection and symptoms clinically are variable and even though humans are affected during childhood, the prevalence is highest in second to fourth decades of life. The incubation period
Lavanya, et al.: Hydatid cyst of the buccal mucosa

is highly variable, and their expression of symptoms depends on the location, pressure, and size of the enlarging cyst. Patients usually show generalized urticaria and pruritus, which results from sensitivity caused due to echinococcal antigen from the cyst. Sometimes when there is a rupture of the cyst wall, unexpectedly leads to abscess formation and locally recurrent cysts. In the maxillofacial region, it accounts for only 2% of hydatid infections of the body and these signs and symptoms are not pathognomonic.

However, typically hydatid cyst presents as a solitary, unilocular cyst, and 20–30% of cases may have several cysts in one organ or a single cyst in multiple organs.

The occurrence of the cyst intra-orally has been well-documented. Involvement of the buccal mucosa is rare, and only two other cases have been reported in the literature. The latter occurred in a 6-year-old male and was asymptomatic with no organ involvement. The present case involved a daily wage laborer of low socioeconomic status and a history of association with sheep or animals could not be elicited. Georgopoulos et al. is the first to report a hydatid cyst in a solitary organ without the involvement of liver and lungs, even though, the embryos have passed through these organs. If no primary or hydatid cyst in other sites of the body have been found, the diagnosis of hydatid cyst localized to head and neck is pretty challenging for the clinicians.

Echinococcus originates from Greek meaning “hedgehog berry” descriptive of its gross pathology of the lesion. Echinococcus for their complete life cycle Figure 6 requires two mammalian hosts. It has a definitive host (e.g., dog) which port the adult worms and the intermediate host (sheep, pig, human etc.) that harbor the larval stage in the intestinal tract and organs, respectively. Humans in close contact with dogs and sheep usually acquire this infection by ingesting eggs of E. granulosus through contaminated food. The eggs hatch in the intestinal mucosa and inhabit for about 5–20 months, the larvae break through the mucosa and through portal vein they reach the liver and get trapped in the sinusoids. Here, they develop into metacestodes or oncospheres and subsequently develop into unilocular cysts, 6–10 days after ingestion. Liver being the first filter is the most frequently involved organ (55–70%) followed by the lung (18–35%). As the larvae reach lungs from the liver, they cause hydatid disease or cyst in any site or multiorgan involvement. When these larvae escape the liver and lung barrier, it can get localized in any organ. The larvae form small cysts reaching up to 1–5 cm in diameter/year. A fibrous capsule (pericyst) is formed surrounding these cysts at this stage. A germinative membrane is formed under these capsules containing new larvae. This layer secretes a periodic acid-Schiff positive polysaccharide-protein complex which is acellular laminated layer (exocyst) which forms the true wall of the cyst that can grow up to 2 mm in thickness and also protects the inner germinal layer (endocyst). To keep the endocyst in close contact with the pericyst, the germinal layer produces clear fluid, also brood capsules which release protoscolices. The head of the protoscolex evacinates after the ingestion by the primary host, and the larva is then referred to as scolex/scolices. These brood capsules and scolices together form the “hydatid sand.” As the cyst enlarges, the surface area of the laminated layer also increases and thickens. These laminations are believed to be laid down to provide accommodation for growth and repair. At any stage of the life cycle of hydatid, the
pericyst can get calcified. This calcification is not an indication of the dead cyst. Daughter cysts are thought to develop from scolices and do not produce pericyst of their own as, usually they are in contact with the host tissue.\[10\] Ultrastructurally, the cyst consists of an inner layer called as a germinal layer of cells which is supported by a middle layer, an acellular laminated membrane (cuticular membrane/exocyst) that are together referred to as endocyst. The outer layer is a dense fibrous layer that is a reaction of the host to the parasite called as ectocyst (pericyst). The germinal layer can produce multiple internal daughter cysts by asexual budding.\[4,8]\]

Computed tomography scan, ultrasonic scanning, and magnetic resonance imaging detects the cystic lesion in soft tissue areas and daughter cysts precisely.\[32\] Blood investigations show eosinophilia in 30% of patients. Fine-needle aspiration biopsy may show hooklets, scolices or remnants of the laminated membrane. Serological tests such as ELISA, indirect hemagglutination test, latex agglutination, immunoelectrophoresis, and Casoni skin tests are more sensitive where in, a decrease in the titer indicates resolution and increase in titer indicates recurrence of *Echinococcosis*.\[6\] Chemotherapy is usually followed in lesions that are inaccessible for surgery and patients with multiple organ involvement. Scolicidal agents or a combination of praziquantel, albendazole, and mebendazole are used against *E. granulosus*. Albendazole postoperatively for 1-month is usually suggested according to the WHO guidelines.\[8\] Complete surgical removal of the cyst is the single effective treatment for hydatid cyst. Surgical pads soaked with 1.5% cetrimide and 1.5% of chlorhexidine gluconate can also be used.\[12\] Aspiration may lead to increased potential for acute anaphylaxis and spread of the disease. 0.5% silver nitrate and 20% hypertonic saline solutions can be used to prevent possible acute allergic reactions, secondary cyst formation, and also to inactivate the daughter cysts, scolices already present.\[6\] Preoperative diagnosis of hydatid cyst is mandatory to prevent anaphylaxis or local recurrence. Spillage of the cystic content, presence of daughter cysts, and leftover endocyst in the operated field are few reasons for the recurrence of the cyst.\[4\] The prognosis is excellent in cases treated by removal of cyst totally without rupture. Hydatid disease or cyst is unusual as its sequelae are more important than the mass effect of the cyst, regardless of the immense size attained by the lesion.

**CONCLUSION**

Hydatid cyst is a pseudocyst of head and neck region, considered to be very rare in nonendemic areas and the diagnosis of which is usually missed out. Therefore, it should be included in the different diagnosis while interpreting a single slow growing solid mass or cystic mass affecting the oral cavity. Dentists should be suspicious especially in the countries where hydatid disease is endemic, and patients must undergo a thorough systemic investigation to exclude the multiorgan involvement and the possible life-threatening occurrence with a long-term mandatory follow-up.

**REFERENCES**

1. Saez J, Pinto P, Apter W, Zulantay I. Cystic *Echinococcus* of the tongue leading to diagnosis of multiple localizations. Am J Trop Med Hyg 2001;65:338-40.
2. Bouckaert MM, Raubenheimer EJ, Jacobs FJ. Maxillofacial hydatid cysts. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2000;89:338-42.
3. Scherer P, Mischkowski RA, Seifert H, Ortmann M, Neugebauer J, Scheer M, et al. Solitary hydatid cyst in the mandible: Case report and review of the literature. J Oral Maxillofac Surg 2008;66:1731-6.
4. Katlimis H, Oztqrkcan S, Ozdemir I, Guvenc IS, Ozturun X. Primary hydatid cyst of the neck. Am J Otolaryngol Head Neck Med Surg 2007;28:205-7.
5. Nidal K. Hydatid disease: A review and update. Curr Anaesth Crit Care 2004;15:173-83.
6. Alaoglu H, Uckan S, Oz G, Altinnors N. Maxillofacial hydatid cyst. J Oral Maxillofac Surg 2002;60:454-6.
7. Aydin S, Erdogan BA, Eken M, Keser SH, Altintoprak N. Unusual reason of a neck mass: Secondary hydatid cyst. Nati J Otorhinolaryngol Head Neck Surg 2013;1:25-7.
8. Soylu L, Aydogan LB, Kiroglu M, Kiroglu F, Javidzadeh A, Tuncer I, et al. Hydatid cyst in the head and neck area. Am J Otolaryngol 1995;16:123-5.
9. Usharani A, Deepica G, Aruna S, Kulkarni S, Sai Kamal Kumar G, Balamuralikrishna C. Case reports of hydatid disease. J Otolaryngol Head Neck Surg 2013;1:25-7.
10. Georgopoulous S, Korres S, Riga M, Kouvidou CH, Balatsouras D, Ferekidou E. Hydatid cyst in the duct of the submandibular gland. Int J Oral Maxillofac Surg 2007;36:177-9.
11. Lewall DB. Hydatid disease: Biology, pathology, imaging and classification. Clin Radiol 1998;53:863-74.
12. Geramizadeh B. Unusual locations of the hydatid cyst: A review from iran. Iran J Med Sci 2013;38:2-14.