The Journal of Laryngology & Otology, E6, 1 of 3.

doi:10.1017/S0022215108003757

Online Only Clinical Record

Sublingual hydatid cyst: case report and literature review

K T Jumani, A Ananthamurthy*, B Joy†, R C Nayar

Abstract

Objectives: To demonstrate the importance of detailed clinical analysis in the differential diagnosis of a cyst in the floor of the mouth, and to provide an update on current knowledge and treatment of sublingual hydatid cyst.

Case report: A 23-year-old man presented complaining of a swelling in the midline of the sublingual region, present for four months and progressively increasing in size. Ultrasonography of the neck revealed a well defined, hypoechoic lesion in the sublingual region, containing a calcific focus. Fine needle aspiration cytology showed numerous round to oval structures resembling brood capsules, with scolices and occasional hooklets. T1- and T2-weighted, multiplanar magnetic resonance imaging scans showed a well defined, multiloculated lesion in the sublingual region.

Conclusion: Hydatid disease may present as a slow-growing cyst in the sublingual region. Aspiration cytology should preferably be avoided until radiological imaging studies are complete. A high index of suspicion is necessary to diagnose hydatid disease in an unusual location.

Key words: Hydatid Disease; Neck; Sublingual Region

Introduction

Cestodes, or tapeworms, are segmented worms which primarily infest dogs and other canine species. Grazing animals such as sheep, cattle, pigs, horses and camels are intermediate hosts in the life cycle of these worms. Occasionally, humans ingest food or water contaminated with tapeworm eggs and become intermediate hosts, leading to echinococcosis or hydatid disease.[1,2]

The literature contains anecdotal reports of hydatid cysts in the head and neck region. We report a case of hydatid cyst located in the sublingual region, and present a review of relevant literature.

Case report

A 23-year-old man presented to the ENT services at St John's Medical College Hospital, Bangalore, a tertiary referral hospital in South India. He had a four-month history of a swelling in the midline of the sublingual region, which had been progressively increasing in size.

On examination, the swelling was oval in shape, approximately 4 × 3 cms in size, nontender, soft and fluctuant. The oral mucosa was free from the swelling and there were no signs of inflammation (Figure 1). On neck examination, there was no swelling in the submental region. On abdominal examination, neither the liver nor the spleen was enlarged. The diagnoses considered were ranula, sublingual dermoid and lymphangioma.

Ultrasonography of the neck revealed a well defined, hypoechoic lesion in the sublingual region, measuring 5.3 × 3 × 3 cms, with an approximate volume of 29 ml and containing a calcific focus.

Fine needle aspiration of cyst contents was performed via the sublingual route, yielding scant, whitish fluid. Smears were prepared from the aspirate and stained with haematoxylin and eosin, Papanicolaou and May-Grunwald-Giemsa stains. The smears showed numerous round to oval structures resembling brood capsules, with scolices and occasional hooklets. A few acellular membranous fragments were also seen. The surrounding area showed a minimal inflammatory cell response. A diagnosis of a parasitic cyst, most probably hydatid, was made.

T1- and T2-weighted, multiplanar magnetic resonance imaging (MRI) showed a well defined, multiloculated, 5.5 × 3 × 3 cm lesion in the sublingual region. The lesion showed a hypointense rim on all imaging sequences (Figure 3). The contents appeared iso- to mildly hyperintense on T1-weighted images and hypo-intense on T2-weighted images (Figure 3). These imaging features were diagnostic of hydatid cyst.

Surgical excision of the cyst was planned. However, as the patient was unwilling to undergo surgery, he was commenced on oral albendazole 400 mg twice daily for three months. The swelling regressed partially in response to this treatment. After three months, the patient was lost to follow up.

Discussion

Echinococcus granulosus (canine tapeworm) is the causative parasite of hydatid disease. Its life cycle involves two hosts. The primary host (dogs and other canine species) is defined as one in which the adult stage lives or in which the sexual mode of reproduction takes place. The intermediate host (other grazers) is the organism in which the larval stage of the parasite lives or in which asexual multiplication takes place.[1,2] The oncosphere is the fully developed egg, and contains an embryo with six hooklets (hexacanth embryo). When humans ingest food

From the Departments of Otorhinolaryngology, *Pathology and †Radiology, St John's Medical College and Hospital, St John's National Academy of Health Sciences, Bangalore, India.

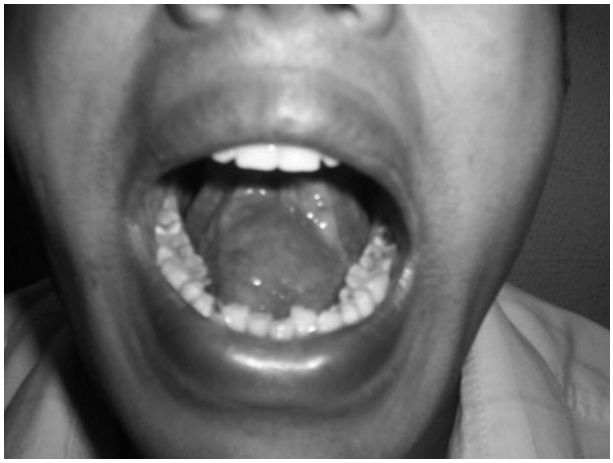

FIG. 1
Clinical photograph of the swelling in the sublingual region.

or water contaminated with oncospheres, they become an intermediate host of the tapeworm.

After ingestion, the parasite emerges from the cyst, penetrates the small intestine wall, and is carried to the liver and other organs via the bloodstream. The larvae lodge within capillaries of various organs and invoke an inflammatory response of mononuclear cells and eosinophils. While many of the larvae are destroyed, a few survive by forming a slow-growing, thick-walled cyst inside which they divide.[3] However, the life cycle, i.e. formation of the adult tapeworm, cannot be completed unless one of the canine species, its primary host, ingests these cysts.

The echinococcus cyst is usually unilocular; however, up to 30 per cent may be multilocular. Cysts may be present in the same organ or in multiple organs,[4] particularly the liver and lungs, occasionally the central nervous system (CNS) and heart, and rarely the musculoskeletal system. In the CNS and musculoskeletal systems, cysts may grow primarily from direct implantation of oncospheres or secondarily from metastatic dissemination of visceral cysts from the liver or lungs.[5] The cyst has a wall comprising three layers. The outermost layer, called the pericyst, is composed of compressed host tissue. The intermediate layer, termed the laminar layer, is derived from the parasite. The innermost layer, termed the germinal layer, comprises the live cellular parasite tissue and is responsible for cyst growth and production of protoscolices, brood capsules (i.e. budded-off pieces of germinal layer with attached protoscolices) and daughter cysts.[6]

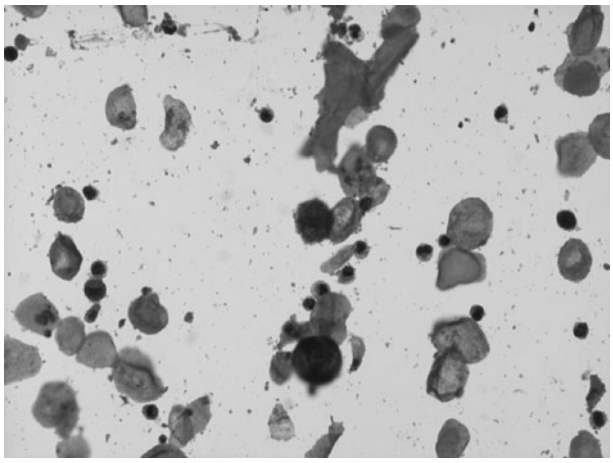

FIG. 2
Photomicrograph of fine needle aspiration cytology smear, showing hydatid scolices (H&E; ×400).

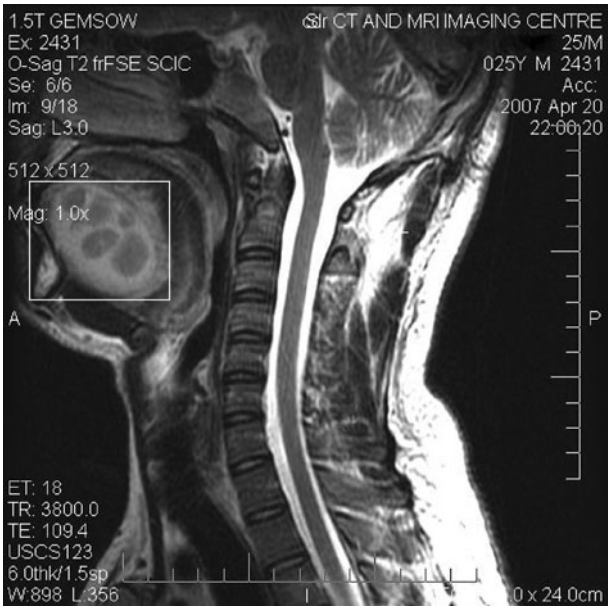

FIG. 3
Sagittal, T2-weighted magnetic resonance imaging scan, showing multiloculated lesion in the sublingual region with a hypointense rim. Note that the contents appear iso- to mildly hyper-intense.

Parasitic diseases are rarely seen in the head and neck region. In a 1998 review by Prousalidis *et al.* of 49 Greek patients with hydatid cysts located in various organs other than the liver and lungs, none was located in the cervical region.[7] Primary hydatid cysts located in the sublingual region are extremely rare even in endemic areas, and very few cases have been reported to date.

The diagnosis of hydatid disease may be established on the basis of clinical presentation, radiography and imaging techniques such as ultrasonography (US), computed tomography (CT) and MRI. Laboratory tests, the intradermal Casoni's test and serological examinations have a limited role in diagnosis as these have poorer diagnostic sensitivity and specificity.[7] If spillage of the cyst contents occurs in the pre- or peri-operative periods, the lesion is very likely to transform into untreatable, multiple hydatidosis.[8] Thus, pre-operative diagnosis of hydatid disease is of great clinical significance.

Ultrasonography and CT examination may demonstrate internal septae and daughter cysts.[8–10] Hydatid cysts have certain characteristic MRI features. The cyst wall appears hypointense on all sequences due to its collagen-rich pericyst.[11] The daughter cyst contents appear hypointense on T1-weighted images, compared with the parent cyst, and hyperintense on T2-weighted images.[12–14] However, the imaging features depend on the stage of the disease. Signal intensity may change with coexistent infection, calcification or haemorrhage.[12,14]

In the present case, the disease was of a long duration and may have been secondarily infected, giving rise to turbidity of the fluid contents and causing the iso- to hyperintensity seen on T1-weighted images and hypo-intensity seen on T2-weighted images.

https://doi.org/10.1017/S0022215108003757 Published online by Cambridge University Press

There are numerous reports of hydatid cysts occurring in unusual sites in the body, often first diagnosed on fine needle aspiration biopsy.[15–18] Fine needle aspiration biopsy in hydatid disease is potentially risky, as spillage of embryos causes contamination and possible fatal anaphylactic reaction; however, the pathologist may encounter hydatid cysts, with typical cytomorphological features, in unexpected sites. Hence, a high index of suspicion is warranted.

The surgical management of a hydatid cyst consists of removal of the cystic material, including the germinative layer, and subsequent obliteration of the potential space occupied by the cyst; this is referred to as a cystotomy and capitonnage.[6] If the germinative layer is left behind, it could lead not only to recurrence but also to suppuration of the cyst cavity. Cystectomy or so-called enucleation is an alternative surgical procedure.[6,19]

Alternative therapies advocated in the management of patients with recurrence or high risk of contamination include puncture, aspiration, injection and reaspiration with non-toxic scolicidal agents (such as 20 per cent hypertonic saline, 0.5 per cent silver nitrate, 95 per cent sterile ethanol, absolute alcohol and mebendazole 2.4 μg/ml),[20,21] or combination chemotherapy using imidazole derivatives, particularly albendazole given orally in a dose of 15 mg/kg (as two divided doses) administered in cycles of 28 days' treatment interrupted by 14 days' rest.[1]

Conclusion

The case of a slow-growing cyst in the sublingual region is presented. Although the diagnosis was suggested by fine needle aspiration, without any complications, aspiration cytology should preferably be avoided if hydatid cyst is suspected until radiological imaging studies are complete, as cyst puncture and escape of hydatid fluid may lead to anaphylactic shock and also to secondary cyst formation.[22] A high index of clinical suspicion is necessary in order to diagnose hydatid disease in unusual locations.

References

1 White AC Jr, Weller PF. Cestodes. In: Kasper DL, Braunwald E, Fauci AS, Hauser SL, Longo DL, Jameson JL, eds. *Harrison's Principles of Internal Medicine*, 16th edn. New York: McGraw-Hill, 2005;**1208**:1272–6
2 Strohl WA, Rouse H, Fisher BD. Protozoa. In: Strohl WA, Rouse H, Fisher BD. *Lippincott's Illustrated Reviews: Microbiology*. Philadelphia: Lippincott Williams & Wilkins, 2001;279–88
3 McAdam AJ, Sharpe AH. Infectious diseases. In: Kumar V, Abbas AK, Fausto N. *Robbins & Cotran Pathologic Basis of Disease*, 7th edn. Philadelphia: Saunders, 2004; 343–414
4 Kammerer WS, Schantz PM. Echinococcal disease. *Infect Dis Clin North Am* 1993;**7**:605–18
5 Nath K, Prabhakar G, Nagar RC. Primary hydatid cyst of neck muscles. *Indian J Pediatr* 2002;**69**:997–8
6 Morris DL, Richards KS. Hydatid disease. Current Medical and Surgical Management. Oxford: Butterworth-Heinemann Ltd, 1992;**3**:43–44
7 Prousalidis J, Tzardinoglou K, Sgouradis L, Katsohis C, Aletras H. Uncommon sites of hydatid disease. *World J Surg* 1998;**22**:17–22
8 Akal M, Kara M. Primary hydatid cyst of the posterior cervical triangle. *J Laryngol Otol* 2002;**116**:153–5
9 Fradis M, Podoshin L, Goldstein Y, Miselevich I, Boss JH. Cervical echinococcal hydatid cyst. *J Laryngol Otol* 1989; **103**:435–7
10 Soylu L, Aydogan LB, Kiroglu M, Javadzadeh A, Tuncer I. Hydatid cyst in the head and neck area. *Am J Otolaryngol* 1995;**16**:123–5
11 Gupta S, Rathi V, Bhargava S. Unilocular primary spinal extradural hydatid cyst. MR appearance. *Indian J Radiol Imaging* 2002;**12**:271–3
12 Singh S, John S. Bilateral adnexal hydatidosis in primary infertility. *Am J Roentgenol* 1999;**173**:1412–13
13 Ozarmagan S, Erbil Y, Barbaros U, Salmaslioglu A, Barboza A. Primary hydatid in the adrenal gland: a case report. *Braz J Infect Dis* 2006;**10**:362–3
14 Singh S, Korah IP, Gibikote SV, Shyam NK, Nair A, Korula A. Sacral hydatidosis: values of MRI in diagnosis. *Skeletal Radiol* 1998;**27**:518–21
15 Das DK, Choudhury U. Hydatid disease: an unusual breast lump. *J Indian Med Assoc* 2002;**100**:327–8
16 Handa U, Mohan H, Ahal S, Mukherjee KK, Dabra A, Lehl SS *et al*. Cytodiagnosis of hydatid disease presenting with Horner's syndrome: a case report. *Acta Cytol* 2001; **45**:784–8
17 Das DK, Bhambhani S, Pant CS. Ultrasound guided fine-needle aspiration cytology: diagnosis of hydatid disease of the abdomen and thorax. *Diagn Cytopathol* 1995;**12**:173–6
18 Giuffre G, Mondello P, Inferrera A, Furchi A, Gentile HM, Speciale G. Unexpected cytological diagnosis of two cases of echinococcosis. *Pathologica* 1993;**85**:747–53
19 Sennaroglu L, Onerci M, Turan E, Sungur A. Infratemporal hydatid cyst – unusual location of echinococcosis. *J Laryngol Otol* 1994;**108**:601–3
20 Taylor DH, Morris DL. The current management of hydatid disease. *Br J Clin Pract* 1988;**42**:401–6
21 Russell RCG, Williams NS, Bulstrode CJK. Parasitic infections. In: Chiodini. *Bailey & Love's Short Practice of Surgery*, 24th edn. London: Arnold, 2004; 146–74
22 Amice J, Sparfel A, Petillon F, Amice V, Jezequel J, Riviere MR. Hydatid cyst of the neck: diagnosis by fine needle aspiration. *Acta Cytol* 1992;**36**:454–6

Address for correspondence:
Dr Kiran T Jumani,
Clinical Research Fellow (SpR),
Dept of Otorhinolaryngology,
The James Cook University Hospital,
Marton Road,
Middlesbrough TS4 3TJ, UK.

Fax: +44 1642854040
E-mail: kiranjumani@gmail.com

Dr K T Jumani takes responsibility for the integrity of the content of the paper.
Competing interests: None declared