Case Report

Amita K*, Rajini T, Sanjay M, Abhishek MG, Prashantha K

Intramuscular hydatid cyst of thigh masquerading as a soft tissue tumour diagnosed by fine needle aspiration cytology

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Abstract: Introduction. Hydatid cyst, caused by *Echinococcus granulosa*, occurs rarely in the musculoskeletal region. Most of the time, clinically and radiologically it is diagnosed as a soft tissue tumor, benign or malignant. There are a few case reports of hydatid cyst presenting as an intramuscular thigh mass, which has been diagnosed at fine needle aspiration cytology (FNAC). Accurate preoperative diagnosis is essential in view of specific therapeutic options for this disease. Here we report a case of hydatid cyst occurring in an unusual location (thigh) and masquerading as a soft tissue tumour, diagnosed at FNAC. Case Report. 56-year-old male patient presented with gradually increasing swelling of the left thigh since 3 years. On examination, there was a firm non-tender 25 × 20 cm swelling on the posterior aspect of left thigh extending from the gluteal region to five cm above the knee joint. An ultrasound diagnosis of a soft tissue tumor was made. At FNAC, fluid was aspirated and smears showed granulomas along with multiple hyaline acellular membrane-like fragments, few showing vague laminations. A diagnosis of hydatid cyst was made at FNAC which was corroborated at histopathology. Conclusion. Intramuscular hydatid cyst of the thigh is a very rare manifestation. The possibility of hydatid cyst should be considered while aspirating any soft tissue mass lesion, especially when fluid is obtained and microscopy shows acellular hyaline membrane-like material, even when fewer laminations are noted.

Keywords: Cytology, Musculoskeletal, *Echinococcus granulosus*, Parasite, Hydatid cyst

1 Introduction

Diagnosis of parasitic infestations by Fine Needle Aspiration Cytology (FNAC) is usually incidental. Case series documenting the role of FNAC in parasitic infestation diagnosis have focussed on superficial palpable nodules, most of which are cysticercosis. [1] Hydatid cyst is most commonly encountered in the liver and its aspiration is often feared due to the risk of anaphylactic reaction. Hence the features of hydatid cyst at cytology are not clear and may be confused with the more common cysticercosis. Similarly, hydatid cyst is uncommon in the thigh. [2] Most of the time,it is diagnosed clinically as a soft tissue tumour, benign or malignant. There are few case reports of hydatid cyst presenting as an intramuscular thigh mass, which has been diagnosed at FNAC. Accurate preoperative diagnosis is essential in view of specific therapeutic options for this disease. [3] Here we report a case of hydatid cyst occurring in an unusual location (thigh) diagnosed at FNAC.

2 Case report

A 56-year male presented with swelling in the left thigh of three years duration. The swelling was insidious in onset, gradually progressing. It was associated with pain for six months. He also had a complaint of difficulty in walking for one month. There was no history of trauma at the site of swelling. There was no history of fever, loss of appetite, weight loss or any other significant history. On local examination, vague swelling in the posterior aspect of thigh measuring 14 x 12 x 7 cm was noted. The skin over the

***Corresponding author: Amita K,** Department of Pathology, Adichunchanagiri Institute of Medical Sciences, Adichunchanagiri University, BG Nagara-571448, Mandya, Karnataka, India, email: dramitay@gmail.com
Rajini T, Sanjay M, Abhishek MG, Adichunchanagiri Institute of Medical Sciences, Adichunchanagiri University, BG Nagara-571448, Mandya, Karnataka, India
Prashantha K, Faculty of Natural Sciences, ACU-Centre for Research and Innovation, Adichunchanagiri University, BG Nagara-571448, Mandya, Karnataka, India

swelling was normal. The swelling was not freely mobile, was firm, and appeared prominent on extension of the leg. No inguinal lymphadenopathy was noted. No similar swelling was noted anywhere else in the body. The provisional clinical diagnosis was that of a soft tissue tumour.

On ultrasound examination, the lesion was described as a well-defined homogenous isoechoic lesion with multiple septations and cystic change. The ultrasound diagnosis was concurrent with the clinical diagnosis of a soft tissue tumour.

FNAC was advised. FNAC was done by standard technique using a 22-gauge needle and 10 cc syringe. On aspiration, 0.5 CC of clear fluid with minute whitish membranous material was obtained. Smears were stained with haematoxylin and eosin (H& E) and Giemsa stain. (Figure 1) Smears showed scanty cellularity comprising of predominantly hyaline eosinophilic acellular membranous material. Most of these hyaline membranes were homogenous appearing. Along with this, epithelioid cells in clusters and singles, foreign body giant cells, and histiocytes in a background of debritic material was noted. Even after careful scrutiny of each of the membranous fragments, the nature of this membranous material was not clear. A question as to whether the membrane was of cysticercosis, hydatid or mucin origin remained. Hence to confirm the nature, re-aspiration was performed. Though most of the membranes on repeat smears showed similar appearance as in the previous FNAC smears, occasional fragments showed laminations characteristic of hydatid cyst. Hence a final FNAC diagnosis of a parasitic lesion consistent with hydatid cyst with granulomatous reaction was made.

Local excision was performed and the specimen was sent for histopathological evaluation.

Gross anatomy- A single large grey brown to grey black soft cystic mass measuring 22 x 14.5 x 12 cm was received. The external surface was intact. The cut surface showed a multiloculated cyst with a focal solid area. Thick

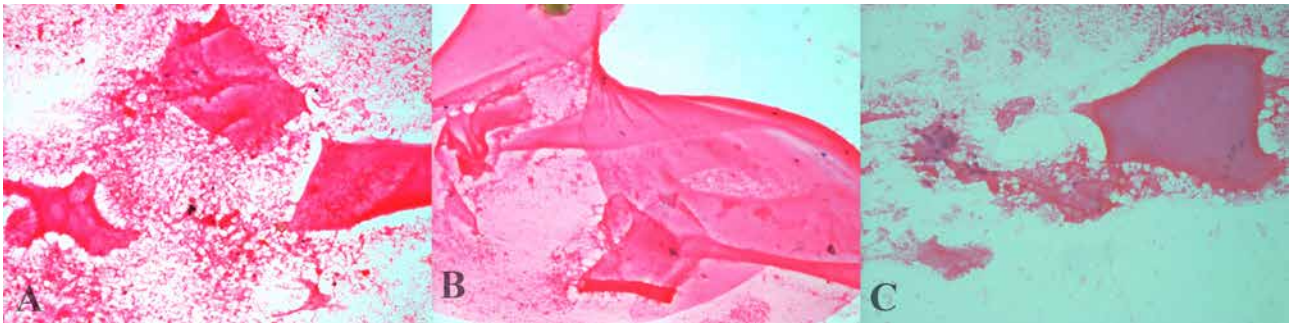

Figure 1A & 1 B. FNAC smear showing granular eosinophilic membrane in a debritic background with few vague laminations (H & E, x200)
Figure 1C. Smear shows membranes adjacent to epithelioid cell clusters. (H & E, x100)

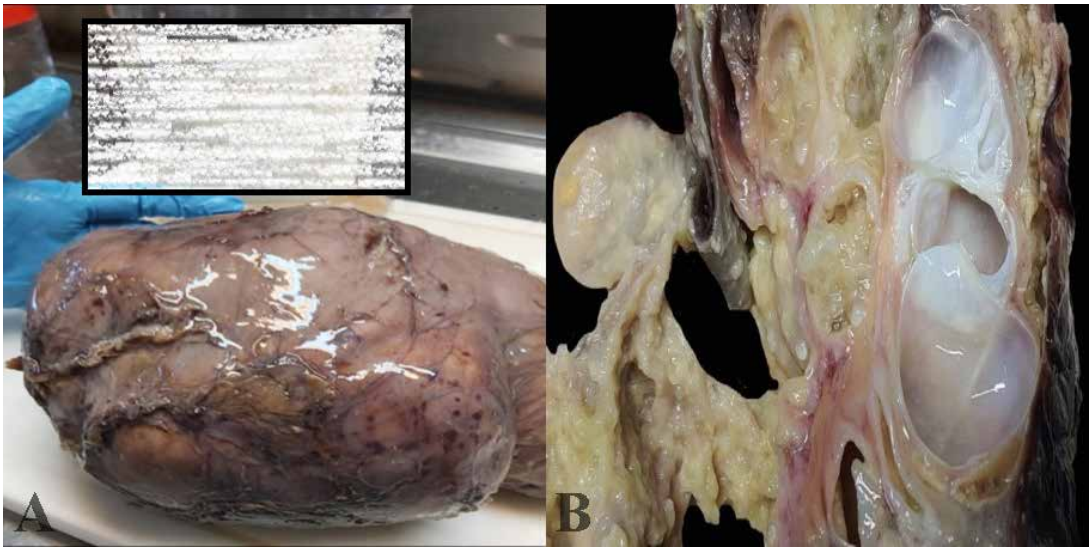

Figure 2 A. Gross specimen showing external surface of intact hydatid cyst with adjacent muscle.
Figure 2 B. Cut surface of cyst showing thin whitish membrane depicting characteristic tender coconut appearance.

grey yellow gelatinous material was expelled out of the cyst. The largest cyst measured 1 cm whilst the smallest cyst was 0.2 cm. The cyst had the typical tender coconut appearance (Figure 2).

Histopathology showed multiple cystic structures with a pericyst comprising of skeletal muscle bundles and granulomatous giant cell reaction. The endocyst consisted of outer fibrous and inner germinal epithelium with few daughter cysts noted; the cyst showed characteristic acellular laminated appearance. (Figure 3 & 4) A final diagnosis of intramuscular hydatid cyst with giant cell reaction was made.

Informed consent: Informed consent has been obtained from all individuals included in this study to perform FNAC and also to utilize the data for publication purposes.

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance with the tenets of the Helsinki Declaration. Ethical committee clearance was exempted for the present work.

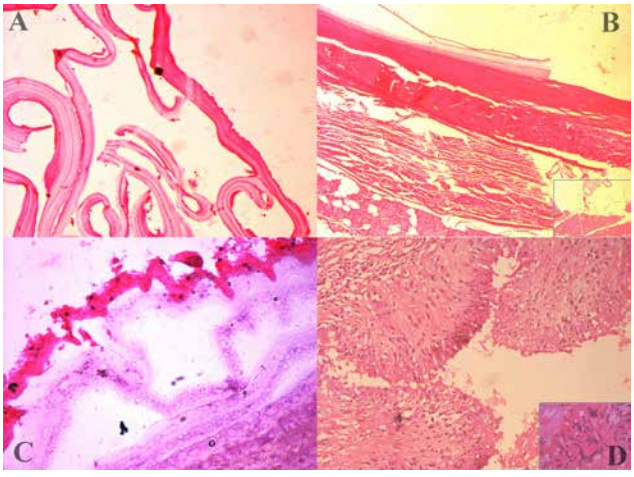

Figure 3A. Section showing laminated membrane. (H & E, x 100)

Figure 3B. Laminated membrane with adjacent skeletal muscle. (H & E, x 100)

Figure 3C. Section shows granular layer of hydatid cyst. (H & E, x 100)

Figure 3D. Section shows pericyst comprising of epithelioid cells and inflammatory cells in the fibrous wall. Inset shows multinucleate giant cells. (H & E, x 100)Figure 1C. Smear shows membranes adjacent to epithelioid cell clusters. (H & E, x100)

3 Discussion

Hydatid cyst (HC) is caused by the larval form of *Echinococcus granulosus*, the adult form of which resides in the intestine of dogs and other canine animals. Human beings are frequently affected in childhood due to common acquittances with dogs. However, the cyst takes a long time to develop and produce symptoms. As a result, hydatid cyst is most commonly diagnosed in middle age or elderly age groups as seen in the present report.

The most common site of HC is the liver (75%) followed by the lung (10%). Rare sites reported in the literature include brain, orbit, omentum, breast, thyroid, tonsils, bone, muscle and soft tissue, each of which not accounting for more than 5%. [4] Prevalence in the musculoskeletal system and soft tissue is rare, around 0.5% to 4% of cases. Intramuscular HC alone accounts for less than 3% of cases. [5,6] The contractile nature and high lactic acid levels makes the muscular tissue hostile for implantation of HC. In the largest series of 272 cases of HC, incidence in the thigh was 0.37%. [7] The existing literature on HC of the thigh is from different parts of the world. Only few cases from India have been reported.

The pathology of primary intramuscular HC of the thigh is not clear. Dissemination of the eggs via lymphatics or direct extension from adjacent sites has been proposed as the mechanism, though the exact cause still remains obscure. Among the reported cases of HC of the thigh in India, most are secondary cases which had occurred due to direct entrance from the surrounding tissue; prevalence of primary HC is rare.

Clinically, most HC are asymptomatic. Frequently, HC is discovered during the investigation for some other disease or it may present with vague symptoms like mass, pain etc. [8] Clinical presentation of musculoskeletal HC is characterized by the presence of a mass lesion which is often confused with a soft tissue tumor, hemangioma, abscess or malignancy.[9] In a soft tissue location, a diagnosis of sarcoma is not a surprise. Rupture of the cyst may lead to a secondary inflammatory response to the released cyst content. Other symptoms reported include limitation

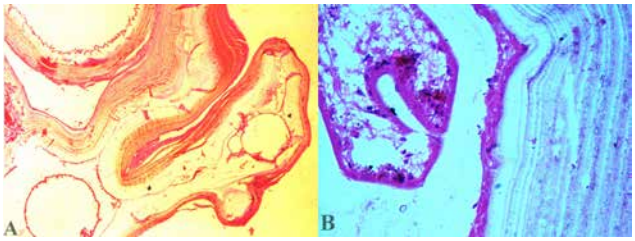

Figure 4A & B. Section shows daughter cyst. (H & E, x 100)

of movements and vessel impingement leading to vascular insufficiency. In a study of 55 cases of abdominal HC, 34% were asymptomatic and 40% were symptomatic. [9]

Radiology is not a reliable tool in the accurate diagnosis of HC in all cases. The lesions show variable appearances on CT or MRI appearing as isodense, hyperdense, homogenous or heterogenous and are commonly mistaken for an abscess or a neoplastic process.[10] Though few authors have described specific findings of HC at MRI, the accessibility and the cost incurred using MRI makes it less suitable in the rural set up of a developing country. Even serology, though positive in many cases, shows false negative results in the presence of senescent or dead cysts. [11] Similarly, cysts in the musculoskeletal region show negative serology. Rickard MD et al, reported a high false negative rate of 20% for serology in diagnosis of HC. [12]

Cytologic techniques have been used for the diagnosis of various palpable lesions since the 18 th century. In mass lesions of the soft tissue and musculoskeletal region, the identification of the nature of the lesion provides crucial information to the clinicians in planning management. With increasing experience and use of radiologic guidance, precise localization and better sampling has been possible. Despite this, the aspiration technique and interpretation require considerable skills and dexterity. There appears to be a general agreement that this method has high specificity and sensitivity; however variations exist depending upon the site and the nature of the lesion.

The risk of dissemination of a daughter cyst during surgery is high when initial preoperative diagnosis is missing. [9] Accurate and precise preoperative diagnosis of HC is important from the therapeutic standpoint as well as of prognostic value. Knowledge and awareness of the rare sites of occurrence of hydatid cyst and its cytomorphological features are essential prerequisites to avoid missing the lesion at FNAC. The accuracy and safety of FNAC in diagnosis of HC in liver is well established. Nevertheless, performing FNAC in suspected cases of HC is fraught with risk in view of anticipated anaphylaxis and spread. However, its usefulness in diagnosing skeletal HC has not been assessed [13.14].

The cyst of HC has two walls, the outer ectocyst which is an acellular thick membrane, beneath which lies the endocyst made predominantly of non-cellular thin germinal epithelium layer. This germinal epithelium gives rise to brood capsules and the membrane of the scolices. Each HC may develop into daughter cysts, each of which have brood capsules and many scolices. In long standing cases, the ectocyst is surrounded by a pericyst representing the host's reaction to the cyst. The pericyst comprises of inflammatory cells rich in eosinophils, epithelioid cells, giant cells and fibrovascular proliferation. At times calcification may be noted.

At cytology, depending upon the site of aspiration and the duration of the cyst, various combinations of features can be seen. Cytology smears in HC show the morphology of the three layers, the frequency of the presence of which may vary from case to case. The most common element seen in the smear is the laminated cyst wall. Ideally, protoscolex, a germinal layer and hooklets can be seen. [15] Protoscolex can be identified in smears by its pyriform shape measuring 300 micrometers and its attached hooklets. The hooklets are small dagger-shaped refractile structures, dispersed in the background. Hooklets are difficult to identify in smears rich in macrophages. Rarely calcification of hooklets may occur, which may be misinterpreted as psammoma bodies. In the present study, the presence of eosinophilic membranes with only a few showing laminations made the interpretation challenging. Granulomas and an inflammatory response were noted in the present study. Most of the studies in the literature on FNAC of HC report similar findings. At times only laminated membrane and inflammatory cells are noted, making the diagnosis challenging. However, with an appropriate clinical picture, substantiated by radiologic findings, a diagnosis of hydatid cyst can still be rendered. [16] Likewise, only granular debris and hooklets may be seen in the smear. Some experts have described the presence of hooklets and scolices as pathognomonic of HC at FNAC. [14-16] In a report by Das et al, accurate diagnosis of HC at FNAC was possible in only 5 out of 8 cases.[13] In contrast, in a study by Tareq et al involving 55 cases, 44 cases were diagnosed accurately by ultrasonography (USG) , while in 11 cases, USG guided FNAC provided accurate diagnosis.[8] In the present study, at FNAC 0.5cc of clear fluid was aspirated. Meshram et al mentioned in their report that the clear fluid may provide a clue to the presence of HC [17].

Another common caveat is to differentiate HC from cysticercosis, with which it shows some common features like the presence of hooklets, scolices and membrane. However, careful attention to the cytology may avoid this error. In cysticercosis, the membrane is thin and nucleated, as compared to the thick laminated membrane of HC. Additionally, the scolices of cysticercosis are large (1mm) and single, as compared to the multiple small oval scolices of HC. Moreover, the hooklets in cysticercosis are large (130-170 micrometers), as compared to their smaller size (22–44 micronmeters) in HC [17].

Excision of an intact cyst with surrounding tissue is the main treatment and avoids recurrence. Nevertheless, instillation of Albendazole into the cyst after aspiration or surgery inactivates the scolices, thereby avoiding recur-

rence. The duration of chemotherapy with solicidal agents depends on the size, location and type of surgery. Praziquantel is added as a combination drug, with the rationale of preventing encystment of protoscoleces following perioperative spillage [8].

4 Conclusion

Intramuscular hydatid cyst of the thigh is a very rare manifestation. The possibility of hydatid cyst should be considered while aspirating any soft tissue mass lesion, especially when fluid is obtained and microscopy shows acellular hyaline membrane like material, even when fewer laminations are noted.

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