Unusual Location of Primary Hydatid Cyst: Soft Tissue Mass in the nape of the Neck and Review of Literature

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ABSTRACT

Hydatid cyst is a zoonotic infection endemic in several regions including the Middle East, Mediterranean countries, South America and South East Asia. Lung and Liver constitute the most common sites of hydatid cyst. Rarely, however, primary hydatid cyst may be found in other parts of the body including the musculoskeletal system presenting with a picture of a slow growing benign cystic swelling. We report a case of a primary hydatid cyst in the neck in a 25 year female who underwent complete excision of the cyst in Pt. B. D. Sharma PGIMS Rohtak.

INTRODUCTION

Hydatid disease is an infestation caused by the parasites of the genus Echinococcus and they are endemic in the temperate regions of the world such as Middle eastern countries, Mediterranean Countries, South America and South East Asia. [1,2] Most commonly found in the liver and lung, they are rarely found in other sites as well, with the clinical picture resembling a slow growing benign tumor. [3][4][5]

CASE REPORT

A 25 year old female presented to General Surgery OPD in Pt. B. D. Sharma PGIMS, Rohtak with chief complaints of a painful swelling on the nape of her neck on the left side since 4 years. The swelling was gradually increasing in size. Patient had no history of fever or any other chronic illness. There was no history of jaundice. Upon physical examination a smooth cystic swelling was noted on the nape of neck on the left side measuring approximately 9x8 cm. The swelling was non-tender and mobile. Swelling was not transilluminant. The patient was further worked up with a diagnosis of benign cystic swelling. (Fig 1)

Ultasonographic examination of the swelling showed a thick walled, multiloculated lesion measuring 7.5 x 8 cm. No focal lesions were seen in the liver. Lateral X ray of the neck showed no bony involvement. No focal lesions were seen on the chest X ray. Contrast enhanced computed tomography of the neck showed a large multiloculated cystic lesion in left paraspinal muscles extending from the level of angle of mandible to the scapula. Multiple daughter cysts were noted towards the inferior margins of the swelling. Tissue surrounding the swelling was comparatively hyperdense with minimal peripheral enhancement. No intraspinal extension was noted.

MRI with T1 Spin Echo and T2 Weighted Fast Spin Echo showed a multiloculated cystic smooth space occupying lesion with thin wall calcifications in the left paraspinal space, displacing the paraspinous muscles and extending from C2 to D3 vertebral level. Fluid aspirated from the swelling was clear and paucicellular. Fluid examination was inconclusive. Patient was planned for complete excision of the cyst with a diagnosis of primary hydatid cyst.

Complete surgical excision of the cystic lesions without spillage was performed. The gross specimen consisted of a soft, smooth walled cyst measuring nearly 7 x 8 cm. (Fig 3) Microscopic examination showed a multiloculated cyst containing clear fluid and multiple daughter cysts. Wall of the cyst contained several scoleces and the diagnosis of hydatid cyst was confirmed. Patient was started on Albendazole 400 mg twice daily for 4 weeks. At 6 months follow up the patient was doing well with no fresh complaints.
DISCUSSION

Hydatid cyst is an infestation caused by the parasites of the order Cestoda, commonly known as flatworms and family taenia more commonly known as tapeworms. Two of the most common species involved as Echinococcus granulosus and Echinococcus Multilocularis. [6]

The adult worms measure nearly 5mm in length and reside in the small intestine of carnivores most commonly dogs and they serve as the definitive host for the organism. [7] Eggs of the organism are shed in the stools of these animals, which are then ingested by the secondary host or intermediate hosts such as cows, sheep, buffaloes, zebra, etc. Humans are incidental host to the organism and typically exposure occurs by ingestion of contaminated vegetables or meat, which is not adequately washed. [8] In the duodenum, the onchospheres hatch and become activated releasing the embryo which produces lytic secretions to enter the portal and lymphatic circulation of the host. [9]

These embryo are then lodged mostly in the hepatic sinusoids and sometimes in the pulmonary capillary bed, and therefore liver (75%) and lungs (15%) are the most commonly affected organs. [3] Upon reaching appropriate site the post-oncospheral development occurs with emergence of metacestode stage. Within 4-7 days the cystic development is completed with the formation of vesicle with germinal layer. [9] Isolated hydatid cyst in other organs have also been reported such as the musculoskeletal system (1-4%), bones, brain, etc. [2] Spleen is the most common extra-hepatic and extra-pulmonary site. It is postulated that such a spread occurs via the lymphatic route. [7] The overall incidence of extra-hepatic and extra-pulmonary hydatidosis has been reported to be 9%. [10]

Isolated hydatid cyst in extra-hepatic and extra-pulmonary sites often remain asymptomatic much like a slow growing benign tumor usually growing at the rate of 1-5 cm a year. [9][11] Symptoms are usually due to pressure over the surrounding structures often when the cysts are more than 5 cm in size. [11] Echinococcus multilocularis is more aggressive than E. granulosus and it may even mimic a malignancy. [12]

Diagnosis is usually made on the basis of history, clinical examination, imaging and serology and typically the individual is often from a rural background, presenting with a slow growing swelling. Physical examination is typical of a soft tissue cystic swelling.

It may be difficult to diagnose an isolated hydatid cyst in an unusual location with imaging even in endemic regions. [12] Ultrasonography usually shows membranes, septations and daughter cysts within the cavity. [7] Increasingly CT and MRI have been found to be more helpful. CT scan typically shows floating membranes and hydatid sand in a cystic lesion. [12] It may also show detachment of membrane of the cyst as linear areas of increased attenuation often referred to as ‘Water Lily sign’. [12] CT scan is also helpful in defining the relation of the cyst with the surrounding structures particularly when defining the relation with osseous structures as well as demonstrating the calcifications in the peripheral rim of the cyst. [12] MRI, although used less often, can be a valuable investigation for complex cysts in head and neck region with more solid component. [11][13][14]

The usual image is that of a cystic lesion with multiple vesicles or ‘bunch of grapes’ appearance, with a peripheral hypointense rim on T2 weighed images or ‘rim sign’. [12] Separate low intensity linear floating structures may also be seen which represent the separated collapsed membranes of the daughter cyst or ‘serpent sign’. [12] Imaging, however, may not be able to conclusively diagnose the condition and thus making a pathological diagnosis important.

Several serological investigations such as direct hemagglutination, latex agglutination and immunoelectrophoresis are available. Serological tests however are not sensitive and specific, therefore while a positive test might prove useful, a negative test does not necessarily rule out the diagnosis of hydatid disease. [13]

Imaging investigations should also be done to evaluate for the presence of a pulmonary or hepatic lesion. Once the diagnosis is confirmed patient may be given Albendazole treatment preoperatively to reduce the size of the swelling. [14] Following which patient should be excised in toto. Extra care should be taken to avoid spillage of any contents since it may cause a severe anaphylactic shock. If the cyst cannot be excised in toto, then PAIR technique can be used wherein, the fluid contents are aspirated, and then irrigated with scolicidal agents such as hypertonic saline. [16] Following removal the patient should be continued on Albendazole. [14]
Fig 1 : Image showing swelling over the nape of neck.

Fig 2 : CECT images showing a multiloculated smooth cystic swelling with no evidence of intraspinal involvement.

Fig 3 : Intraop-image showing a smooth cystic lesion in the paraspinal space
CONCLUSION

Currently, the most effective treatment for hydatid disease located in soft tissue is still surgery. The main purpose of surgery is to prevent complications such as compression of surrounding structures, infection, or cyst rupture. Total cystectomy with fibrous adventitia, which allows for the removal of all parasitic elements without spillage of the contents of the cyst, is curative treatment for soft tissue hydatidosis. Soft tissue cysts can be easily ruptured. Therefore, rupture of the cyst must be avoided to prevent recurrence. Based on our own experience, regardless of the location, all patients with hydatid cyst for which surgery is planned should be given albendazole prophylactically 400 mg/d for at least 2 weeks. No postoperative medical treatment is needed for patients who had total cystectomy. In cases with cyst rupture or with other cysts in different locations, treatment should be continued for 2 months during the postoperative period.

REFERENCES