

## A Rare entity of Primary Splenic Hydatidosis – A case report

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### **ABSTRACT**

Hydatid disease is endemic in farming areas but can occur worldwide. It is caused by infection with *Echinococcus granulosus* leading to development of cysts. The most common site of disease is liver and lungs. Other sites such as spleen, pancreas, heart and muscles are rarely affected. Isolated splenic hydatidosis is a rare entity and only small clinical series or case reports have addressed the issue of splenic echinococcosis. We hereby report a case of giant hydatid cyst in spleen of 39 year old female. Splenectomy was done and diagnosis was confirmed on histopathological examination. Thus hydatid disease should be kept in mind while dealing with cystic lesions of spleen.

**KEYWORDS:** Hydatid disease, Echinococcosis, Spleen.

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### **INTRODUCTION**

Hydatid disease (Echinococcosis) is a zoonotic infection caused by the larval form of parasites of tapeworm, *Echinococcus granulosus*. Humans are the accidental intermediate host in the development cycle of hydatid disease. It is an endemic disease in the sheep and cattle raising countries Middle East, North Africa, New Zealand, Australia, and South America. Although hydatid disease affects any organ or soft tissue, it most frequently found in

liver (60–70%), lungs (30%), and rarely encountered in the kidney, spleen, bone, thyroid, breast and pancreas. Clinical presentation varies according to the anatomic location of the cyst. Usually, splenic hydatid cysts are secondary, either resulting from spontaneous spread of cysts or occurring after operations involving hydatidosis in other regions.<sup>1</sup> We report a case of a primary splenic hydatid cyst treated surgically.

**CASE REPORT**

A 39 year old female patient came with complaints of left upper abdomen pain since one and a half years. The pain was continuous, dull aching with a dragging sensation with lump in abdomen since 2 months. Her vital parameters were normal. On physical examination, abdomen was distended with a tender, palpable mass in the left hypochondriac region. There was no lymphadenopathy, rebound tenderness, guarding or hepatomegaly. The chest, cardiovascular, central nervous and the musculoskeletal systems were normal on examination. The routine laboratory investigation CBC, coagulation profile, biochemistry, renal function test, liver

function test and electrolytes revealed no abnormalities. Serological test for echinococcus was positive. (Echinococcus - Ig G serum – 6.9)Chest X ray was normal. The abdominal USG showed multiloculated cystic lesion in the left hypochondriac region. Abdominal CT scan showed multiple, complex, cystic hypodense lesions in spleen. (Figure 1) The cysts contained multiple hypodense daughter cysts within. There is rim of wall calcification along inferior aspects of larger cyst. The patient received pneumococcal vaccine two weeks prior to surgery. Exploratory laparotomy was done and spleen was removed.



Figure 1: CT Abdomen showing complex cystic hypodense lesion in spleen.

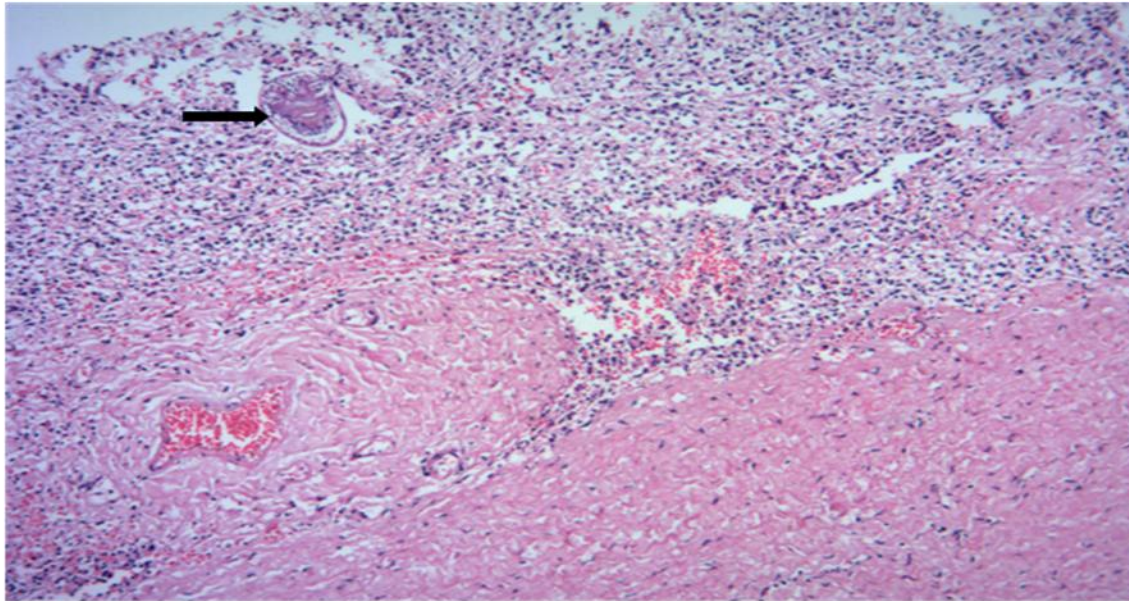
Grossly enlarged spleen measured 17x13x5cms and weighed 1060gms. The external surface is nodular and mottled with grayish white areas. On cut surface, multiloculated cysts were seen. The cysts contained multiple, variable sized daughter cysts. (Figure 2)



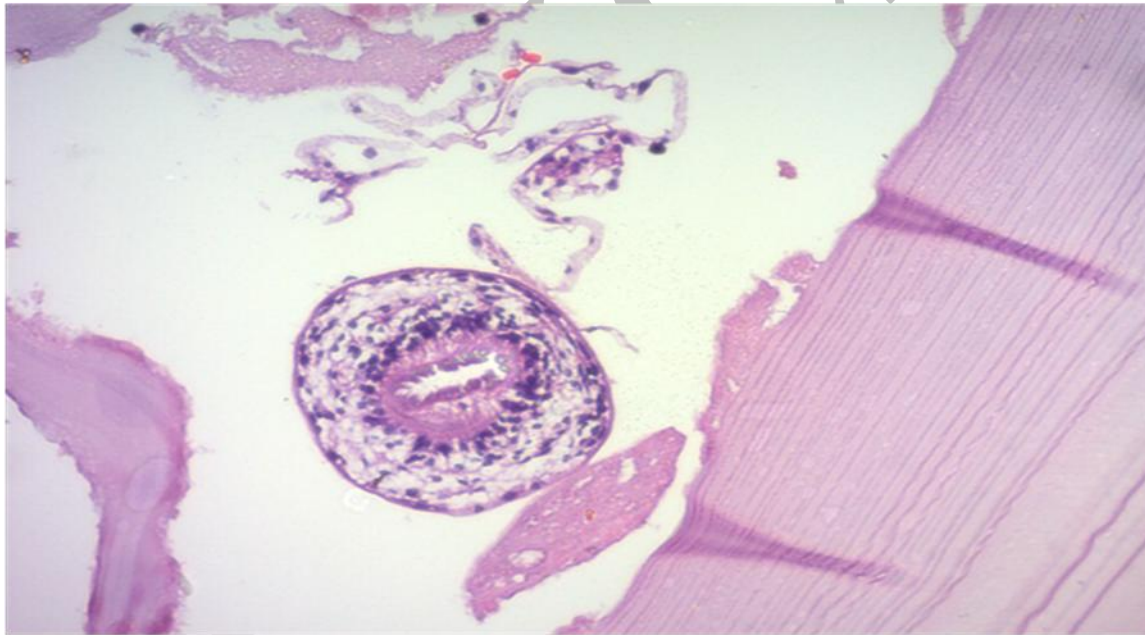
Figure 2: Grossly enlarged spleen: Cut surface showing multilocular cyst filled with daughter cysts.

Microscopic examination revealed an outer laminated acellular chitinous ectocyst, inner germinal endocyst with numerous scolices. (Figures 3 & 4) The acellular laminated membrane of the echinococcal cyst stained strongly for Periodic-acid Schiff (PAS) stain.

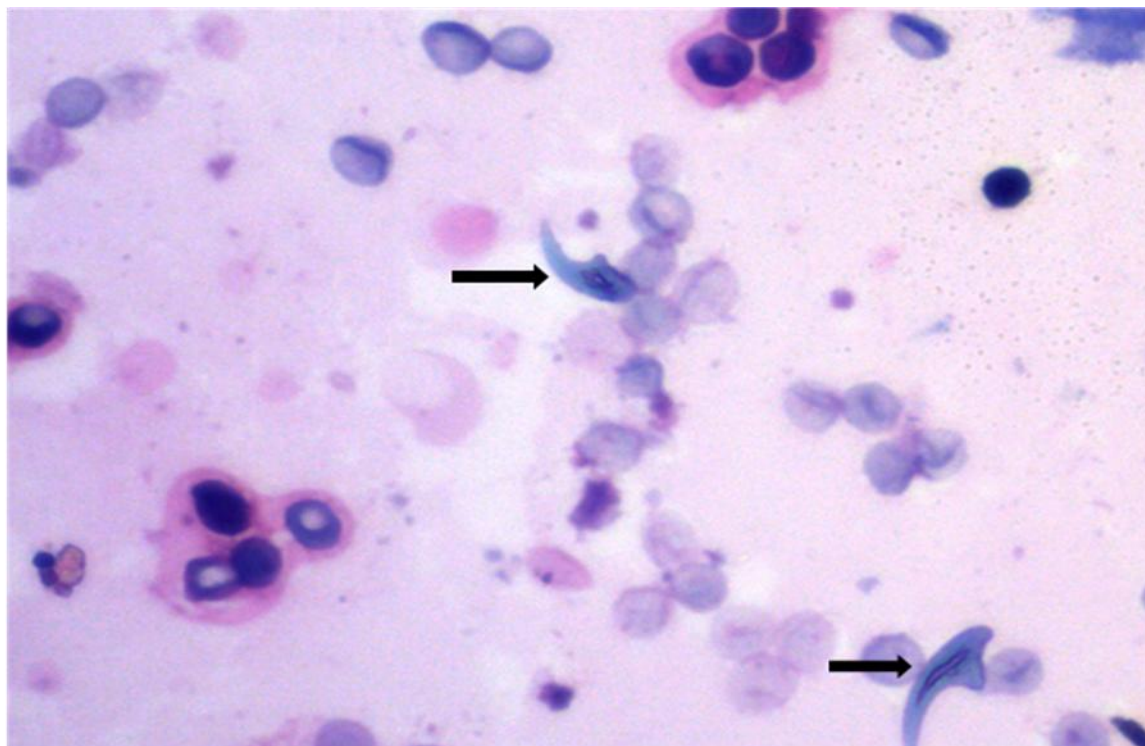




**Figure 3: Scolex within splenic parenchyma. H & E stain. 10X**



**Figure 4: H & E section showing lamellated cyst wall with a scolex. 40X**



**Figure 5: Centrifuged deposits showing hooklets in the aspirated fluid. Giemsa stain**

## **DISCUSSION**

Echinococcus completes its life cycle involving dogs (definitive host), and sheep and goats (intermediate hosts). Humans are the accidental intermediate host and infection occurs through the ingestion of contaminated vegetables, water or soil contaminated by the excreta of infected host or intermediate host. The liver is the first and the main barrier to parasitic embryos which migrate from the intestine and gain access to the portal circulation. The majority of the embryos are trapped in the liver and if the embryos bypass the liver, the lung is the

next most frequently involved organ. Echinococcal cysts may develop anywhere from the toe to the crown of the head. Practically no organ is immune to infestation by echinococcosis. Rare sites include the spleen, thyroid, gall bladder, central nervous system, kidney, psoas sheath, retroperitoneal region, orbit, cervix and adductor longus muscle.<sup>2</sup>

Splenic Echinococcosis is rare in endemic regions accounting for 0.5 to 8% of all cases of echinococcosis.<sup>3</sup> Berlot first described splenic hydatid cyst as an autopsy

finding in 1790. An incidence of 4.3% has been reported in India, with the highest in central India.<sup>4</sup> Splenic echinococcal cysts account for 1.5 to 3.5% of all abdominal echinococcosis and represents nearly two-thirds of all cystic lesions of the spleen.<sup>5,6</sup> Primary infestation of the spleen usually takes place by the arterial route after the parasite has passed the two filters (hepatic and pulmonary). A retrograde venous route, which bypasses the lung and liver, is also reported. Some complications may be seen with splenic hydatid cyst such as secondary infection, fistulisation to adjacent organs and rupture into the peritoneal cavity. The traumatic or spontaneous rupture of a hydatid cyst may cause a life-threatening complication of systemic anaphylactic reaction.<sup>1</sup>

Cystic lesions of the spleen include parasitic cysts, benign neoplastic cysts such as lymphangiomas, cavernous hemangiomas or dermoid cysts, and non-neoplastic cysts (pseudocysts) resulting from haemorrhage or area of infarction. Echinococcal cyst is the only parasitic cyst to affect the spleen and is reported to be twice as common as the non-parasitic variety.<sup>3,8</sup> Hydatid cyst are slow growing and can reach a enormous size and maybe asymptomatic. Splenic cyst presents

with local or referred pain or signs and symptoms related to splenomegaly, abdominal distention and compression of nearby structures.<sup>8</sup>

Sonography and Computed Tomography are the most valuable imaging techniques for diagnosis and evaluation of splenic diseases. Serological test are highly sensitive and specific for echinococcosis.<sup>8</sup> In our case, USG revealed cystic lesion of spleen and serological test was positive for echinococcosis.<sup>8</sup> CT is the best modality to detect calcification and internal cystic structure behind calcification..

On histopathology, the hydatid cyst consists of three layers. The outermost adventitia (pseudocyst) is formed of compressed splenic tissue, a middle layer laminated membrane of friable ectocyst and an innermost germinal layer, endocyst.<sup>8</sup>

The fluid aspirated from hydatid cyst is usually clear and contains debris, a few inflammatory cells and numerous scolices. The finding of hooklets is diagnostic of hydatid disease on cytology.<sup>9</sup> We had aspirated fluid before opening the spleen and the fluid centrifuged showed presence hooklets and few scolices.(Figure:5)

Complications include superadded infection, rupture and fistulisation into



bowel mainly colon. Rupture may lead to anaphylactic reactions, portal hypertension.<sup>10</sup>

Splenectomy is the standard treatment of choice. Recently splenic conservative surgery has emerged as the treatment of choice in suitable cases and also to reduce the incidence of post-splenectomy opportunistic infections.<sup>10</sup>

**CONCLUSION:** Splenic involvement by hydatid cyst is low as compared to other abdominal viscera and should be kept in mind in dealing with cystic lesions of spleen especially where the disease is endemic.

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