Primary splenic echinococcal cyst: a rare presentation

Pinki PANDEY, A DIXIT, S CHANDRA, V CHATURVEDI, Anuradha SHARMA

Departments of Pathology and Pharmacology, U P Rural Institute of Medical Sciences and Research, Saifai, Etawah, India, Department of Pathology, Maharishi Markandeshwar Institute of Medical Sciences and Research, M M University, Haryana, India

ABSTRACT

Human echinococcosis is caused by the larval stage of the tapeworm Echinococcus granulosus. The most commonly affected sites are the liver and lungs. Primary splenic echinococcosis is rare. It may occur in association with hepatic, pulmonary or multi-organ echinococcosis, or very rarely as isolated primary splenic echinococcosis. We report the unusual case of isolated primary splenic echinococcal cyst in a 15-year-old boy who presented with the history of left upper abdominal swelling and pain. Ultrasonography revealed a splenic cystic lesion with numerous daughter cysts suggestive of echinococcosis that was confirmed on computed tomography scan. Radiographic imaging studies did not show any cyst in liver, lungs or kidneys. Splenectomy was performed with an uneventful recovery. The diagnosis of splenic echinococcosis was confirmed on histopathological examination. Spleen, although a rare site for presentation of hydatid disease, should always be kept in mind while dealing with cystic lesions of the spleen especially in endemic regions.

Keywords: Hydatid cyst, parasitic infection, splenic cyst

INTRODUCTION

Echinococcosis is a zoonotic problem of man caused by the larval stage of the cestode of the genus Echinococcus. There are four species and three are of medical importance (E. granulosus, E. multilocularis and E. vogeli). It is endemic in sheep and cattle rearing regions, including the Middle East, Mediterranean countries, Central Europe, South America, Indian subcontinent, Australia, New Zealand and South Africa. The liver, followed by lungs is the most commonly affected sites in adults. Echinococcosis of the spleen is extremely rare even in endemic regions accounting for 0.5 to 8% of all cases of echinococcosis. An incidence of 4.3% has been reported in India, with the highest in central India. 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.

Correspondence author: Pinki PANDEY
Type V, B-301, New campus, UPRIMS&R, Saifai, Etawah. 206301. UP, INDIA
Tel: +919416825149
E mail: pnkdxt@yahoo.co.in

Splenic echinococcal cysts occur at all ages and affect both genders. They may be detected incidentally or may present with non
CASE REPORT

A 15-year-old boy from the rural area of Punjab presented with a four-week history of increasing left upper abdominal swelling, heaviness and pain. Abdominal examination revealed a tender irregular soft mass in the left upper quadrant and lumbar region that moved with respiration. The rest of the examination was normal. His general condition was good and past medical history was otherwise unremarkable. Routine laboratory investigations were within normal limits. Serology for the Human Immune Deficiency Virus (HIV) was negative.

An ultrasound scan of the abdomen revealed a well-defined unilocular cystic lesion with evidence of numerous daughter cysts in the spleen that were confirmed with a computed tomography (CT) scan (Figures 1a and b). Radiographic imaging studies did not show any other cyst in the liver, lungs or kidneys. The patient received pneumococcal vaccine (Pneumovac) two weeks prior to surgery. Exploratory laparotomy through a left subcostal incision revealed a large cyst occupying the spleen. Complete splenectomy was performed without damaging the cyst wall. The splenectomy specimen showed a large cystic mass resembling the albumin of boiled egg of size 12 x 10 x 5cm (Figure 2a). The cyst wall was of uniform thickness and numerous daughter cysts of variable sizes were seen on sectioning. Microscopic examination revealed an outer laminated acellular chitinous ectocyst (Figure 2b), inner germinal endocyst with numerous scolices, and an outermost pericyst formed as a response to host tissue reaction. The acellular laminated membrane of the echinococcal cyst stained strongly for Periodic-acid Schiff (PAS) stain (Figure 2b). The patient was treated with albendazole 400 mg twice daily for four weeks and had an uneventful recovery. At one year follow-up, there was no evidence of relapse.

Figs. 1: a) Ultrasound scan of the abdomen showing splenic hydatid cyst with daughter cysts, and b) an axial contrast-enhanced computed tomography scan showing a huge splenic cyst with a daughter cyst in the superior portion of the mother cyst.
DISCUSSION

Normally *Echinococcus* completes its life cycle involving dogs (definitive host), and sheep and goats (intermediate hosts). Humans are the accidental intermediate host and 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. Practically no organ is immune to infestation by echinococcosis. Rare sites include the spleen, thyroid, gall bladder, central nervous system, kidney, psoas sheet, retroperitoneal region, orbit, cervix and adductor longus muscle.  

*Echinococcal cyst* consists of two layers: an outer, thick, non-nucleated, carbohydrate rich acellular laminated layer (ectocyst), and an inner, nucleated, germinal layer (endocyst) from which brood capsules develop. There is also an adventitial layer or pericyst which is formed by the host reaction. The cyst contains deposits of brood capsules and scoles at the bottom, known as hydatid sand. Daughter cysts are formed when these brood capsules are attached to the germinal layer of the mother cyst by a pedicle.

Cystic lesions of the spleen are rare and 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. Most occur as part of disseminated involvement. Primary splenic involvement is rare as the cyst embryos are trapped in the liver and/ or the lungs after ingestion, and therefore do
not reach the systemic circulation to infect the spleen. Secondary splenic involvement may occur as a result of rupture of a hepatic echinococcal cyst with abdominal and pelvic dissemination. 2 Splenic echinococcal cysts are mostly solitary but multiple cysts have also been reported. 9 The disease affects all age groups and both sexes with equal frequency. 4 The cyst grows at a rate of 0.3 – 1 cm/year and it may take 5-20 years to grow to a sufficient size (3-35cm) to cause symptoms. 10

The clinical presentation depends on the size and site of the cyst. Most patients remain asymptomatic and are discovered incidentally, or may present with non-specific symptoms like left hypochondrial pain, dyspepsia, dyspnoea or painful mass in the left upper quadrant as was noted in our case. Patients can even present with complications such as cysts rupture into the peritoneal cavity, gall bladder, biliary tree, pleural cavity or hepatic veins, or as severe anaphylactic reactions due to rupture of the cyst leading to fever, pruritus, dyspnoea, stridor and oedema of the face. 10

Different serologic tests such as: serum immunoelectrophoresis which is currently the most reliable, with a sensitivity of approximately 90%, ELISA, Latex and indirect haemagglutination assay (IHA) tests. These are helpful for the diagnosis, screening and post-operative follow up for recurrence. 6 Pre-operative diagnosis is mandatory in order to prevent any rupture of cyst so as to avoid anaphylactic shock or local recurrence. This can be made on ultrasound and confirmed by CT scan (94-96% and 100% sensitivity respectively). 11 Marginal or crumpled egg-shell like calcification in the splenic area on the abdominal or chest radiograph are suggestive of splenic echinococcosis. Ultrasonography scan may reveal a solitary unilocular lesion, or rarely, multiple well-defined anechoic spherical cystic lesions with hyperechoic marginal calcification in the spleen. 12

Splenectomy has been considered the treatment of choice. Splenectomy must be carried out in large and giant hydatids of the spleen localised in the organ or in its hilum, and in irreversible derangement of the organ function. However, due to its immunologic significance, conservative therapeutic approaches (spleen-preserving surgery) have been developed for the management of splenic cysts. This achieves removal of the lesion while preserving splenic function, thereby reducing the risk of uncontrolled post-operative infection. Treatment may consist of partial splenectomy, cyst enucleation, deroofing of cyst with omentoplasty or, external drainage. Conservative techniques are used for superficial cysts confined to one of the poles of spleen and cysts with extensive adhesions. Preservation of spleen should always be tried in children to avoid overwhelming post-splenectomy infection (OPS1). 13, 14, 15

With the advent of minimal access surgery, most of these techniques can be performed through laparoscopic surgery. For patients with small-sized, superficially located cysts, laparoscopic techniques yield success rates similar to the conventional open surgery. 17 Polat et al. compared open and laparoscopic hydatid cystectomy and showed that the morbidity rates were 14.2% for laparoscopy and 33% open surgery respectively. The
most frequent post-operative complications with laparoscopic approach were wound infections. 18

Surgery remains the standard treatment, however, pre- and post-operative one month course of Albendazole (10mg/kg/day) and two weeks of Praziquental (50mg/kg/day twice a week) should be considered in order to sterilise the cyst and reduce the chances of anaphylaxis and tension in the cyst wall. This reduces the risk of spillage during surgery and reduces the recurrence rate post operatively. 16 Better forms of chemotherapy and newer methods, such as puncture, aspiration, injection and re-aspiration (PAIR) technique using hypertonic saline or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts. 7

In conclusion, splenic ecinococcosis although rare, should always be kept in mind while dealing with cystic lesions of the spleen, especially in endemic areas like Middle East, the Mediterranean countries, Central Europe, Indian subcontinent, and South Africa. This case is being reported because of its rarity.

REFERENCES