INTRODUCTION

Cystic lesions of the spleen can be either primary or secondary depending upon the underlying etiology. Secondary cysts develop due to hemorrhage, trauma, infarction or inflammation, while the primary cysts may be parasitic or non-parasitic.1 Hydatid cyst due to flat worm Echinococcus causes significant health problem with reasonably high morbidity and wastage of resources and is usually the only parasitic cyst of the spleen.2 It is extremely rare even in the endemic regions with an incidence of 0.5-4%.3 Only limited case reports are published, while isolated perisplenic hydatid cyst is even rarer. One such case is, hereby, reported.

CASE REPORT

A 21-year-old male student presented with 3 months history of gradually increasing discomfort in the left hypochondrium. The pain progressively became moderate to severe and continuous for the last 2 weeks, which interfered with his studies. The pain was aggravated by respiratory movements and associated with nausea. There was no accompanying history of jaundice, fever, abdominal distension or bowel upset. He received symptomatic treatment in the form of analgesics, and reassurance to which, there was no favourable response. He was fond of pets and had recently purchased two puppies. His clinical examination revealed a 4 cm firm tender splenomegaly without any splenic rub. However, there was no jaundice, hepatomegaly, lymphadenopathy or signs of chronic liver disease. His hematological profile showed normal leucocyte count of 7.9x10⁹/L with 16% eosinophils. His liver function tests for serum total bilirubin, ALT and alkaline phosphatase were within normal limits. Ultrasonography of the abdomen showed a cyst in the hilar region of spleen having septations with internal echos. An upper midline laparotomy was performed and a perisplenic cyst was removed along with spleen from the sub-diaphragmatic location. Histopathological examination confirmed acellular fibrous wall of hydatid cyst with germinal layer and scolices in the centre. Postoperatively, patient was continued on oral Albendazole for one month.

ABSTRACT

Splenic parasitic cysts due to flat worm Echinococci resulting in hydatid disease are a rare presentation as primary site even in the endemic regions. Primary splenic parasitic cysts have an incidence of 0.5-4%. A 21-year-old male with pet dogs at home, presented with 3 months history of gradually increasing discomfort in the left hypochondrium and tender splenomegaly. He had marked eosinophilia with normal liver function tests and positive serum IgM Echinococcus antibodies. Ultrasonography showed a cyst in the hilar region of spleen having septations with internal echos. An upper midline laparotomy was performed and a perisplenic cyst was removed along with spleen from the sub-diaphragmatic location. Histopathological examination confirmed acellular fibrous wall of hydatid cyst with germinal layer and scolices in the centre. Postoperatively, patient was continued on oral Albendazole for one month.

Key words: Hydatid cyst. Splenic cyst. Echinococcus.
Hydatid cyst is caused by the larval stages of the genus *Echinococcus* granulosus and is primarily a disease of sheep and cattle. Humans are the accidental intermediate hosts. It occurs predominantly in young adults in developing countries due to bad hygiene and poor water sanitation. It is more prevalent in males with an annual incidence ranging from 1-220 cases/100,000 persons in endemic areas. The typical route of human transmission includes contaminated soil, infected animals and consumption of unwashed vegetables. The ova are ingested via faeco-oral route. The cyst capsule is dissolved in the acidic environment of the stomach and ova are released which hatch in the duodenum penetrating its wall to enter the portal circulation to reside in the liver. Liver accounts for the most common site for hydatid cyst at 52-77% followed by lungs at 10-40%. Primary or secondary foci can be seen in almost all organs including brain, kidneys, bones and spleen.

Clinical presentation depends upon the site and size of the lesion. Asymptomatic cysts may persist for years growing at 0.3-1 cm/year. However, when the cysts reach an advanced size, the patient complains of symptoms due to pressure effects. Pain in the left hypochondrium is the classical presentation in cases of splenic hydatid cyst. Infected cysts with abscess formation are accompanied by fever and leucocytosis. Dramatic clinical picture is seen when there is rupture into the peritoneal space resulting in anaphylactic reaction.

Pre-operative diagnosis is made on ultrasound and CT scan with a greater specificity and sensitivity. Ultrasound shows solitary, unilocular or rarely multiple well-defined aneohogenic, spherical cystic lesions. A double contour view of the endo- and pericyst is highly diagnostic. Complicated cyst appears as having air fluid level, water lily sign, meniscus sign and cavity. CT scan of the abdomen confirms the cystic lesion with or without daughter cysts in the spleen with an attenuation value near that of water not enhancing after contrast administration. Ancillary investigations may show eosinophilia of greater than 7% in about 30% patients. Different serological tests are carried out for diagnosis, screening and postoperative follow-up. The most commonly used serological tests are Indirect Hemagglutination Assay (IHA) test and Complement Fixation Test (CFT). The CFT becomes negative and is important in postoperative follow-up. Immunoelectrophoresis is the most important serological test specific for *Echinococcus* granulosus and reverts to negative after successful eradication and treatment of the cyst.

Histological examination of the cyst wall shows an outer chitinous or fibrous laminar layer and an inner germinal layer. The cyst wall is surrounded by granulation tissue or a fibrous capsule called pericyst layer. Calcification signifies that the cyst is dead, while daughter cysts and brood capsules with scolices are seen in the viable cyst.

The mode of patient management either conservatively or surgically is still debatable. Conservative treatment is a common practice in endemic areas. Medical treatment with anti-helminthics is used as a prophylaxis before carrying out ultrasound guided cyst decompression or surgery. Anti-helminthics like praziquantel, mebendazole and albendazole are used but Albendazole is preferred because of its protoscolicidal effects which prevent seeding subsequent to accidental spillage. There is 30% resolve response rate with 30-50% decrease in the size of the cyst. The treatment is continued upto one month postoperatively as well.

Open splenectomy is the standard surgical procedure. Enucleation, pericystectomy and simple cystotomy with captonnage are parenchyma saving procedures currently practiced by most surgeons. Partial splenectomy was introduced for trauma and benign splenic diseases after recognition of fatal post-splenectomy sepsis. Laparoscopic approach has also been advocated for uncomplicated hydatid cyst as a safe and effective option with favourable long-term results. Risk of spontaneous rupture followed by anaphylactic reaction is an absolute indication for surgical management.

To conclude, primary splenic involvement is an uncommon manifestation of hydatid disease and should be suspected on detection of any splenic cyst in an endemic areas such as ours. Public health measures and awareness is the most effective tool for control of hydatid disease to prevent morbidity and drainage of resources.
REFERENCES


