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CASE REPORT

ISOLATED GIANT ECHINOCOCCOSIS IN SPLEEN

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ABSTRACT

Echinococcosis is a zoonosis caused by the larval stage of cestodes of the genus *Echinococcus*. The tiny worm lives in the intestines of dogs, their ova pass on to sheep, through feco-oral route, and the larvae come to rest in sheep organs, develop into metacestodes (cysts loaded with protoscoleces). When these organs loaded with protoscoleces are ingested by dogs, the life-cycle is completed and repeats. Humans are accidental intermediate hosts. The most commonly involved organs are liver (75%), lung (15.4%) and spleen (5.1%). The cyst grows slowly and may take 5-20 years to grow into a size to cause symptoms. In 90% of cases *Echinococcus granulosus* involves a single organ and of these 70% have a single cyst. Usually splenic echinococcal infection is the result of spontaneous rupture and spread of cysts from the liver. Rarely the larvae bypass the liver and lung and reach other organs like the spleen. Spontaneous or traumatic rupture of a hydatid cyst may cause life threatening systemic anaphylaxis and is the main cause of morbidity and mortality in this zoonotic disease. This is a report of primary Echinococcal infection of the spleen, causing massive splenic enlargement without involvement of any other organ. The patient was treated by splenectomy ensuring no spillage of cyst contents despite its about to rupture status. The case report and discussion outline the management protocol for isolated echinococcosis of the spleen.

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INTRODUCTION

This 40-year-old lady had been symptomatic for the previous three months with backache. Evaluation by a physician a month ago revealed a left hypochondrial lump, which was clinically consistent with splenic enlargement. There were no symptoms or signs suggestive of hepatic decompensation or local inflammation. Ultrasonography confirmed splenic enlargement due to a large cyst with multiple daughter cysts. CT Scan confirmed gross enlargement of the spleen with a span of 24.8 cm due to a large cyst within the spleen measuring 17.8 cm x 14.4 cm x 11 cm with several septations suggestive of daughter cysts within. She had severe anemia with Hemoglobin level at 6 gm/dl. Leukocytes and platelets were within normal limits. Hemoglobin electrophoresis did not reveal any abnormality., serum ferritin was also within normal limits. She was treated with Tablet Albendazole 400mg HS for 10 days, transfused two units of packed cells, and was immunized with Pneumovax prior to Splenectomy. Under GA a generous Mercedes Benz shaped incision was taken to visualize the large spleen its vessels and its attachments with minimum handling of the spleen itself.

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At exploration the massive spleen was seen to have several white and discolored patches on its surface. No additional cysts were noted in the liver or peritoneal cavity. The abdominal cavity around the spleen was packed with mops soaked in 10% Povidone Iodine. The short gastric vessels in the gastro splenic ligament were serially clamped and divided to gain access to the lesser sac. The pancreas was identified and the splenic vessels were ligated in continuity. The spleen was then gently delivered into the wound to visualize the lienorenal ligament, which too was clamped ligated and divided. The splenic hilum was then addressed and individual vessels in the splenic hilum were clamped ligated and divided to deliver the specimen. Hemostasis was ensured and the splenic bed was washed with 10% povidone iodine. A 24F drain was left and the wound was closed by mass closure technique. Her post-operative recovery has been uneventful and treatment with Albendazole has been restarted and is planned to continue for three months under close supervision. Grossing of the excised specimen revealed several cystic cavities filled with translucent straw-colored fluid and several daughter cysts floating within confirming suspicion of Cystic Echinococcosis of the Spleen. Serial sectioning revealed a large cyst filled with yellowish gelatinous substance and several smaller gelatinous cysts floating freely within the cysts and communicating with multiple smaller cysts towards the periphery. Light microscopic examination revealed a PAS positive, hyalinized cyst wall, focally showing

lamellation. The intracystic contents comprised necrotic, eosinophilic material. Few daughter cysts identified. The gross and microscopic features confirmed Cystic Echinococcosis of Spleen.

DISCUSSION

Echinococcosis is a zoonotic infection caused by the metacestodes (larval stage) of cestodes of the genus *Echinococcus*. The infection is common in sheep rearing regions of the world and is seen world-wide as the definitive host of *Echinococcus* worm is the dog. The disease is endemic in the Mediterranean Countries, the Middle East, New Zealand, Australia, East Africa and South America, the highest incidence being recorded in Iran. The two species of medical importance include *E. granulosus* which causes cystic echinococcosis (CE) as in our case and *E. multilocularis* which causes alveolar echinococcosis (AE). The most commonly involved organs are liver (75%), lung (15.4%) and spleen (5.1%) (Brunetti *et al.*, 2009; Rasheed *et al.*, 2013). Dogs are definitive hosts for the adult *Echinococcus*. The tape worm attaches itself to the villi of the dog ileum and releases thousands of ova in dog feces. Humans become accidental intermediate hosts, through the feco-oral route by the ingestion of food and drink contaminated with dogs' feces or by direct contact with dogs transmitting the ova to the hands and mouth. Following ingestion by the accidental host (humans) or intermediate hosts (sheep, goat, cattle), the oncospheres hatch from the eggs, penetrate the intestinal mucosa, enter the blood and reach the liver, lung, spleen or other organs. In these organs the oncospheres develop into fluid filled cysts which increase in size at the rate of 0.3 cm – 1 cm a year. Multiple layers line the cyst wall forming a metacystode (hydatid cyst), the outermost is the pericyst or fibrous capsule derived from the host tissue.

The hydatid cyst wall itself has two layers, an outer gelatinous laminated layer called ectocyst and an inner germinal layer called endocyst, which secretes hydatid fluid and forms brood capsules. Brood capsules are smaller cysts within the larger cyst in which protoscoleces or future worm heads form. Free floating brood capsules and protoscoleces form hydatid sand. When the organ containing hydatid cyst and protoscoleces containing organ of the intermediate host is eaten by the definitive host, the protoscoleces evaginate, attach to host's intestinal mucosa and develop into adult worms in 30 – 80 days, with proglottid segments producing eggs and the cycle is repeated. Transmission is frequent in settings where domestic dogs eat viscera of slaughtered animals, get infected, pass eggs in their feces which are passed on to other animals or humans via feco oral route. The larval stage of *E. granulosus* causes cystic echinococcosis. The life cycle of *E. multilocularis* involves wild canids as the definitive host and rodents as the intermediate host. Domestic dogs or cats may get infected by ingesting infected rat viscera and may pass on the infection to humans, causing alveolar echinococcosis, *E. vogeli* and *E. oligarthrus* are extremely rare causes of human echinococcosis. Simultaneous involvement of liver, lung and spleen may be seen in 20% to 30% of all cases. Splenic hydatid cysts are usually solitary but multiple cysts may be seen in 25% to 30% of cases (Brunetti *et al.*, 2009; Rasheed *et al.*, 2013; Pukar, 2013). The largest cyst reported has been from Saudi Arabia, containing 37 liters of fluid (Rasheed *et al.*, 2013; Largest Hydatid Cyst, 2005). Spontaneous or traumatic rupture of a hydatid cyst may cause life threatening systemic anaphylaxis

and is the main cause of morbidity and mortality in this zoonotic disease. Hydatid cysts can die with degeneration of the membranes, development of cystic vacuoles and calcification of the wall. Calcification of the wall does not always imply that the cyst is dead. asymptomatic. The cyst grows slowly and may take 5-20 years to grow into a size to cause symptoms. In 90% of cases *Echinococcus granulosus* involves a single organ and of these 70% have a single cyst (Brunetti *et al.*, 2009; Rasheed *et al.*, 2013). Usually splenic echinococcal infection is the result of spontaneous rupture and spread of echinococcal cysts from the liver. Primary isolated splenic echinococcosis is likely through the arterial route if the cestode larvae bypass the first two filters, the liver and lung on their way from the intestine. Rarely it may spread in a retrograde manner from the liver to the spleen via the hepatic portal and splenic veins in portal hypertension or directly from colonic veins near the splenic flexure communicating with the splenic veins (Brunetti *et al.*, 2009; Rasheed *et al.*, 2013; Pukar, 2013). Patients with splenic hydatid cysts are generally asymptomatic. When the cyst advances in size the patient may present with abdominal pain, usually a dull dragging ache. There may be dyspepsia, constipation, due to pressure on the colon, dyspnea due to pressure on the diaphragm, or cough and hemoptysis due to fistulation into the bronchial tree. Occasionally spontaneous or traumatic rupture may present with signs of hemorrhagic or anaphylactic shock.

The usual first indication of splenic echinococcosis is an accidentally discovered mass in the left hypochondrium. Splenic hydatid cysts are usually solitary. As the cyst increases in size it may kink or compress hilar vessels leading to atrophy of splenic tissue and eventually the cyst may replace the entire splenic tissue. Chronic pericystic inflammation may cause adhesions with adjacent organs and even fistulation between the cyst and adjacent organs like stomach, kidney, colon, and bronchus. The imaging findings are similar to hepatic hydatid disease and may range from purely cystic lesions to completely solid appearance. The differential diagnosis of cystic lesions in the spleen include epidermoid cysts, pseudocysts, abscesses, hematomas, lipomas and liposarcomas. Type I, Primary or true cysts are characterized by an epithelial or an endothelial lining of their inner wall and may be parasitic caused mainly by parasites of the genus *Echinococcus* or non-parasitic. Type II, Secondary or pseudocysts do not have an epithelial lining. Splenic pseudocysts are the result of past injury, infection or infarction. Most splenic cysts are asymptomatic and diagnosed incidentally. Large cysts may cause pain and heaviness in the left hypochondrial region, due to distension of the splenic capsule. Symptoms secondary to pressure on surrounding viscera such as nausea, vomiting, and flatulence may also appear. Pressure on the diaphragm may cause pleuritic pain, cough or dyspnea. During clinical examination if incidentally a splenic enlargement is detected, other diseases associated with splenomegaly like portal hypertension, chronic malaria, hemolytic anemia, lymphoproliferative disorders and collagen disorders need to be excluded. Ultrasonography can reliably distinguish between cystic and solid lesions but are not specific even if typical findings of solitary anechoic lesions are demonstrated. In the early stages detection of cystic lesions with daughter cysts, hydatid membranes and hydatid sand may help clinch the diagnosis with a sensitivity of 90-95%. When the cyst contains membranes, mixed echoes may confuse with abscess or neoplasm. When daughter cysts are present, internal septations are seen. Hydatid sand reflects the presence of scoleces from the protoscolices. This may be better appreciated

when shifting the patient's position during imaging. Infoldings of the inner cyst wall, separation of the hydatid membrane from the wall of the cyst and hydatid sand are suggestive of hydatid cyst diagnosis. When the fluid pressure within the cyst is high, dissections may occur and these undulating membranes seen on sonography are known as "Snake Sign". In more advanced stage of collapse, these membranes may appear twisted giving a "Water Lily" appearance. On axial sonograms parallel stripes called "Double Lines" delineate the space between the parasite's ectocyst and host's pericyst. After medical therapy the appearance of "Snake Sign" indicates that the parasite is responding to treatment. Ultrasound can also distinguish between active and transitional and inactive cysts. Signs which suggest that the parasite is dead and the lesion is inactive include an elliptical cyst, a germinal layer which is detached from the cyst wall, cyst fluid returning coarse echoes and calcifications noted in the cyst wall. The WHO-IWGE Classification of Echinococcal Cysts based on imaging gives some idea of the cyst status and is helpful in planning treatment. CE 1 are unilocular anechoic cystic lesions with double line sign seen in active lesions. CE2 are multiseptated, rosette like or honeycomb cyst as seen in our case and is also indicative of an active lesion. CE3A are cysts with detached membranes "Water Lily" sign and CE 3B are cysts with daughter cysts in solid matrix are indicative of cysts in transitional stage. CE 4 are cysts with heterogeneous hypoechoic/ hyperechoic contents, with no daughter cysts are indicative of inactive cysts. CE 5 are solid cysts with calcified walls and these too are indicative of Inactive cysts. CT has a higher sensitivity than ultrasound, (95-100%). CT is best for determining the site, size and number of the cysts. CT is better than Sonography at detecting cysts located at extrahepatic sites.

The attenuation values noted on CT scan depends on the density of cyst contents. Hydatid cysts usually show attenuation values similar to water but high attenuation values may be seen if there is intracystic debris, hydatid sand and pus. Calcification in the wall of the cyst is best seen on non-contrast enhanced CT Scans and is indicative of parasite death. Calcification in the cyst wall and presence of daughter cysts is suggestive of Echinococcal infection. MRI may be useful in patients with negative serology and indeterminate Sonography and CT scan images. Hydatid cysts show a low signal intensity rim the "Rim Sign" characteristic of Echinococcosis best seen in T2 weighted images. T1 weighted images show a cystic mass with a hypointense fibrous pericyst. The hydatid matrix has intermediate signal intensity and peripheral daughter cysts are hypointense relative to the matrix. On T2 weighted images, the matrix is hyperintense and the daughter cysts are relatively hypointense. After pharmacotherapy T2 weighted images shows degenerated cysts with detached membranes seen as "Snake Sign" (Brunetti *et al.*, 2009; Rasheed *et al.*, 2013; Pukar, 2013; Stojkovic *et al.*, 2012). Among the serological methods, immunoelectrophoretic has a 90-95% sensitivity. It remains positive for 1 year after the organism has been eradicated. Indirect hemagglutination has a sensitivity of 85% and remains positive for many years. Since Capron *et al* reported the presence of an antigen specific for *E. granulosus* that appeared electrophoretically as a band of characteristic morphology and location when tested against sera from human patients. They named this band arc 5 because of its relative position in the immunoelectrophoretic pattern. In residual or highly calcified hydatid cysts the arc may be absent. The physical status of the hydatid cyst influences the degree of antigen stimulation of the immune system of the host and the result of the test. False

positive serologic tests have been reported in malignant disease, collagen disorders, hepatic cirrhosis, schistosomiasis, cysticercosis and other parasitic infections. Elevated levels of IgE are nonspecific indicators of active infection / sensitization, whereas elevated levels of Ig M class specific to echinococcal organisms may be sensitive indicators of recurrent disease. Personal history, presence of calcification in cyst wall, daughter cysts, concurrent cystic lesions in liver and / or lungs are helpful in the diagnosis of splenic echinococcosis (Brunetti *et al.*, 2009; Rasheed *et al.*, 2013). Although antihelminthic drugs are used before and after surgery, they cannot replace surgery. Most antihelminthic drugs are absorbed poorly and do not reach adequate concentrations within the cyst cavity to reliably kill the parasites. Several treatment modalities are available including, surgery, percutaneous aspiration injection reaspiration (PAIR), and pharmacotherapy with benzimidazoles (BMZ), but selected modality needs to be tailored to each individual patient. Percutaneous Aspiration Irrigation and Reaspiration (PAIR) can be applied for conservation of the spleen in patients who do not give consent for surgery or are at high risk for anesthesia. It is contraindicated in uncooperative patients, infected abscessed cysts and mature calcified cysts. PAIR + BMZ give the best results in > 5 cm CE1 and CE3A cysts. Results of PAIR are not as good for CE2, CE3B, CE4 and CE5 cysts. Currently, splenectomy is the treatment of choice for isolated splenic echinococcosis, especially larger cysts due to the elevated risk of rupture in larger cysts. Splenectomy however, carries enhanced risk of hemorrhage, pancreatitis, gastric injuries and overwhelming post splenectomy infection. Sepsis related deaths account for 1.9% of adults and 4% of children who undergo splenectomy. The reported incidence of OPSI varies from 0.9% to 60% with a mortality exceeding 50%. Pneumococcus, Hib and Meningococcus C conjugate vaccines should be given two weeks before or two weeks after splenectomy. Spleen preserving surgery like partial splenectomy and cyst enucleation and cyst deroofting with omentoplasty should be considered in children who are at particularly higher risk for OPSI but the splenic parenchyma is compressed and non-functional especially in larger cysts and the thick fibrous pericyst seen in hepatic hydatid cysts is not seen in the spleen making conservative surgery difficult.

The combination of PAIR and BMZ can be effective and safe, but the duration of treatment and follow up is long. Treatment with PAIR is less successful for cysts that appear solid on sonography and for multiseptated cysts that contain multiple daughter cysts, and these are better treated surgically. Multiple cysts, multiple initial locations, recurrence in multiple organs, especially in the peritoneum are an indication for pharmacotherapy. Albendazole is significantly more effective than mebendazole. It is usually preferred at an average daily dose of 15mg/kg/day. It must be given continuously started one month before surgery and continued for three months after surgery. Blood count and transaminases must be checked every week for the first month and every month thereafter. Praziquantel which is protoscolicidal may be added especially after surgery when the risk of inadvertent spillage is perceived to be high. In Alveolar echinococcosis benzimidazoles are started 1-3 months before surgery and for up to 24 months after surgery. In inoperable cases and cases with residual disease benzimidazoles should be continued throughout life. Drugs however cannot be used alone as they have only parasitostatic action and can never effect cure. Intraoperatively the use of scollicidal agents like hypertonic saline, cetrime, alcohol and

0.5% silver nitrate solution injected into the cyst, before opening the cavity tends to kill the daughter cysts and prevent intraperitoneal spread¹. Recurrence is a major problem with all treatment modalities ranging from 4.6% to 22%. The main reason for recurrence is microscopic spillage of live parasite, failure to remove all viable cysts, and part of cyst wall being left behind during surgery. Recurrence is never seen following complete resection. Recurrence may occur many years after so called complete resection hence follow up has to be prolonged and continued as long as feasible. Serologic tests are noncontributory towards diagnosis of recurrence as these titers may remain high in the post-operative period. Recurrence has to be confirmed by USG, CT scan MRI or PET CT scan (Brunetti *et al.*, 2009; Stojkovic *et al.*, 2012).

Conclusion

Echinococcosis of the spleen is a rare disorder. The distinction between parasitic and non-parasitic cysts of spleen is difficult. Sonography, CT Scan and serology can provide some indication but cannot always confirm the diagnosis. A high index of suspicion for echinococcosis has to be kept in mind especially in endemic areas when faced with clinically suspicious signs and imaging. The presence of daughter cysts in a large cystic lesion and calcification of cyst wall are pathognomonic signs. In isolated splenic echinococcosis total splenectomy is the treatment of choice as it offers complete cure. In children spleen preserving surgery should be considered to prevent OPSI.

To prevent recurrence due to inadvertent microscopic and macroscopic spillage during surgery, packing surrounding viscera with hypertonic saline or 10% povidone iodine-soaked mops and preoperative and postoperative pharmacotherapy with benzimidazoles should be ensured.

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