Case Report

Rare Case Of Calcified Congenital Splenic Cyst

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Keywords: True cyst, epithelial lining, calcification, splenectomy

Abstract:-
Splenic cysts are rarely encountered in day to day practice, with less than 1000 cases reported in medical literature. They are classified into true and false cysts depending on the presence of an epithelial lining. Here, we report a case of a giant congenital epithelial splenic cyst in a 49 year old lady with an unusual presentation

Background:-
Splenic cysts are rare lesions. Among them true congenital cysts occur in about 0.07% of patients. The first case was reported by Berthelot in 1790 and since then only about 1000 cases have been reported. These cysts can undergo secondary changes like calcification. However this phenomenon is more common in hydatid cysts and false cysts. Calcification in a true cyst occurs less commonly and only a very few cases have been reported. Here we report a case of a giant congenital splenic cyst with calcification and ossification, which is rarely encountered.

Case Presentation:-
A 49 year old lady presented with pain in the left side of chest associated with left hypochondriac pain since 1 week, with history of fever since 3 days. There was no history of breathlessness, cough, no history of vomiting. She also gave no history of trauma or contact with animals.

On examination, she was febrile with temperature of 101°F, with mild pallor. She had dull note to percussion in the left 1A and 1S areas, with reduced breath sounds in the same lung fields. Abdomen examination was remarkable except for mild tenderness in the left hypochondrium. Clinically a working diagnosis of left pleural effusion was made.
However her CXR showed left pleural effusion with slight shift of cardiac to right side, with a well defined rounded opacity below the left dome of diaphragm with a calcified wall. USG abdomen was done which showed a well defined cyst with calcified wall measuring 10.1x7.9 cm in the left hypochondrium. For further confirmation CT Scan was done, it showed that the spleen was normal in size, with a large cyst measuring 10.51x8.56x9.73 cm in diameters with peripheral egg shell calcification, interposed between splenic cavity, inferior surface of left lobe of liver and greater curvature of stomach, exophytic from splenic hilum with a confluent satellite lesion with uncalcified wall in the upper pole of spleen. It was associated with left sided pleural effusion with sub pleural consolidation. Her routine hematologic and biochemical tests showed Hb of 10.6 g% and ESR was 120 mm/hr, other tests were normal. She was started on IV antibiotics and other supportive therapy.

Since there was suspicion of an infected cyst, staged treatment was planned. Patient underwent Laparoscopic aspiration of cyst contents with deroofing of the cyst wall. Pneumococcal vaccination was administered as splenectomy was planned. Biopsy of the cyst wall showed an epithelial splenic cyst with inflammatory contents, it was negative for evidence of parasite, granuloma or neoplastic pathology.

However she presented within 3 weeks with left shoulder pain and this time repeat CT showed reaccumulation of the cyst, hence she underwent total splenectomy. Her post operative recovery was good and patient was discharged.

Discussion:
Splenic cysts are classified by Martin into true or primary cysts which are characterized by an epithelial lining and false or secondary cysts which do not have one. True cysts can be parasitic or non parasitic. Non-parasitic cysts are further classified into congenital or neoplastic. False cysts are usually secondary to blunt abdominal trauma and are more common than true cysts accounting for about 75% of cases.

Congenital epithelial cysts are rare lesions. Their origin is not clear, it has been proposed that they are derived from inclusion of mesothelial lining into parenchyma during splenic development. They remain asymptomatic for a long period of time or may present with non specific symptoms such as nausea, vomiting, left hypochondriac pain, due to pressure on surrounding structures, making clinical diagnosis difficult. Sometimes they may present with complications which include rupture, hemorrhage or infection which may be life threatening.

Management depends on the patient’s symptoms and size of the cyst. Observation is recommended for asymptomatic patient's and cysts measuring less than 5 cms. Operative intervention is required if the size is more than 5 cms or the patient is symptomatic. In earlier days total splenectomy was performed, but nowadays spleen conserving procedures are being undertaken due to vital functions of the spleen. They include partial splenectomy, cystectomy, cyst decompression.

Laparoscopic technique for decompression of splenic cyst was first demonstrated by Salky et al in 1985. However total splenectomy still remains the procedure of
choice if cyst involves whole of splenic parenchyma or is situated in the splenic hilum\textsuperscript{8}. Our patient had a huge cyst exophytic from the splenic hilum, hence she underwent total splenectomy.

**Conclusion:**
True giant splenic cysts are rare lesions. The pathogenesis and classification still remain unclear and are a matter of debate. Although diagnosis has become easy with vast resources of diagnostic modalities, no clear cut guidelines exist for diagnosing and management of these lesions and are awaited for.

**Figure legends:**
Figure 1: Chest X ray PA view showing left pleural effusion with a well-defined round opacity below the left dome of diaphragm with a calcified wall.

Figure 2 and 3: section of CT abdomen Showing a cyst with peripheral egg shell calcification lying between left lobe of liver and stomach on the right and splenic hilum on the left. A satellite lesion is seen in the upper pole of spleen.
Figure 4: Histopathology examination showing cyst with an epithelial lining consisting of cuboidal cells and inflammation of its luminal contents.

Figure 5: gross specimen Gross specimen of spleen demonstrating the splenic cavity
Figure 6: gross specimen of spleen. Specimen consists of ruptured enlarged spleen measuring 17 X 11 X 8 cms. Open cut section shows irregular cystic cavity which is calcified and ossified.

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