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Case Report

Epidermoid cyst in a wandering spleen: a rare case report

Karthik Chalamalasetti*

Department of Surgery, Kurnool Medical college and Government General Hospital, Kurnool, Andhra Pradesh, India

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***Correspondence:**
Dr. Karthik Chalamalasetti,
E-mail: dockarthikc@gmail.com

ABSTRACT

Epidermoid cyst is a rare disease of the spleen. Primary true cysts account for 10% of non-parasitic cysts. Wandering spleen is a rare finding which is due to failure to form normal peritoneal attachments. Here we discuss a rare case of an epidermoid cyst in a wandering spleen. A 64-year-old female patient presented with abdominal discomfort in the left lumbar region for two months. On examination, an enlarged spleen was noted, which was freely mobile in all directions. CECT abdomen showed a calcified focal cystic lesion in the spleen. The patient underwent laparotomy that revealed a wandering spleen with a cystic lesion involving most of the spleen. Histopathology was consistent with epidermoid cyst of spleen. Epidermoid cysts have a variable presentation and are difficult to diagnose preoperatively and its association with wandering spleen is scarcely reported in the literature.

Keywords: Epidermoid cyst, Spleen, Wandering, Splenic cysts

INTRODUCTION

Epidermal cysts of the spleen are true cysts of the spleen that have an epithelial lining. They are congenital, commonly sporadic, that occur due to genetic defect in mesothelial migration. The epithelial lining is formed from the entrapped peritoneal mesothelial cells during development.[1] Wandering spleen is another rare entity that is caused due to laxity of splenic supporting ligaments. The long splenic pedicle is prone to torsion and splenic infarction.[2] Here we report a rare case of an epidermoid cyst in wandering spleen.

CASE REPORT

A 64-year-old female presented with discomfort in her left upper abdomen for two months. The pain was continuous, dull aching type with increased intensity over that last two weeks. She denies any history of fever, vomiting, trauma or altered bowel habits. On examination, there was an enlarged spleen extending to the level of the umbilicus. In addition, spleen was freely mobile in all directions. Routine blood workup was unremarkable, and an ultrasound of abdomen showed an enlarged spleen with an anechoic cystic lesion. CECT abdomen revealed moderate splenomegaly with a calcified cystic lesion of size 8×6 cm (Figure 1).

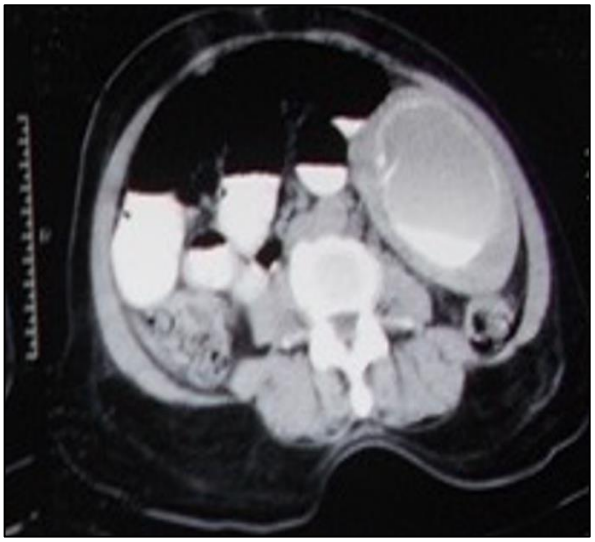

Figure 1: CECT of the abdomen showing cystic lesion in the spleen with wall calcification.

Figure 2: Post splenectomy specimen.

Figure 3: Gross pathology.

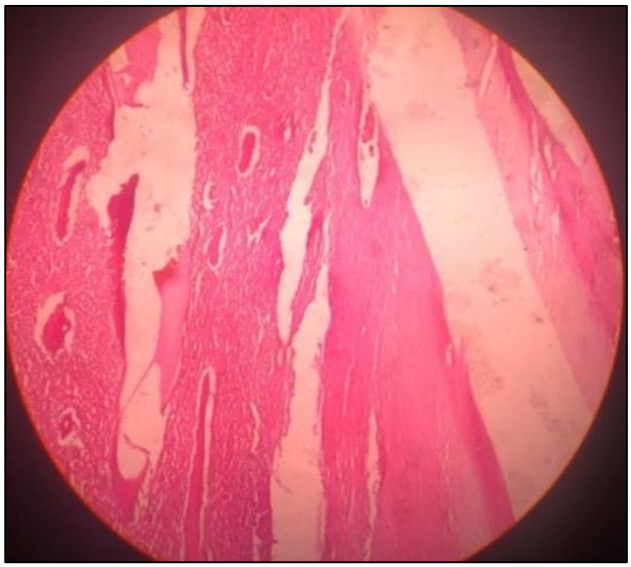

Figure 4: Histopathology.

The patient underwent surgical exploration and intraoperatively there was a cystic lesion involving the cranial aspect of the spleen (Figure 2).

The spleen was freely mobile in the abdominal cavity with an abnormally long splenic pedicle, suggestive of wandering spleen. A splenectomy was done, and the specimen was sent for histopathology. Grossly, a large cyst was noted replacing almost the entire spleen (Figure 3). Histopathological examination was consistent with epidermoid cyst of spleen (Figure 4). Postoperative period was uneventful, and the patient was discharged on postoperative day 5. The patient is on regular follow-up and one year after surgery, the patient is symptom free.

DISCUSSION

Epidermal cysts of the spleen are rare primary cystic lesions lined by stratified squamous epithelial lining.[3,4] These cysts account for 10% of benign non-parasitic cysts occurring in the spleen. These are true cysts owing to epidermal lining in contrast to false, that occur as a result of trauma or haemorrhage.[5] Splenic cysts can be classified into type 1 (true cysts) and type 2 (false cysts). Type 1 cysts can be further classified into congenital and neoplastic or parasitic and non-parasitic cysts. Epidermoid and dermoid cysts constitute congenital cysts, whereas hemangiomas and lymphangiomas are classified under neoplastic aetiology.[6] Around 1/3rd of the splenic cysts are asymptomatic and identified incidentally.[7]

Epidermoid cysts of the spleen appear as anechoic or hypoechoic on ultrasound, may contain internal echoes (debris) or echogenic wall with distal shadowing (wall calcification).[8] A CT scan may show a well-defined thin-walled hypodense cyst, with or without wall enhancement.[1] Hydatid cysts of the spleen are the most common differential diagnosis, especially in endemic areas. Though hydatid cysts contain cyst wall calcification similarly to epidermoid cysts, the clinical presentation is different, and hydatid cysts may contain septations, hydatid sand or lamellar membranes. Another distinguishing feature from hydatid cysts is that they are hyperechoic, whereas epidermoid cysts are anechoic or hypoechoic. Other important differential diagnosis includes lymphangiomas and pseudocysts.[9]

Partial or total splenectomy is the treatment of choice.[3,10] Other management options such as aspiration or incision and drainage have also been described. Partial splenectomy may be indicated in smaller cysts (less than 5 cm), whereas total splenectomy may be required for larger cysts. Spleen preservation surgery is preferred whenever indicated to avoid post-splenectomy life-threatening complications. However, total splenectomy may be preferred in large cysts to prevent complications such as haemorrhage and rupture.[11]

Wandering spleen is a congenital disorder where spleen is freely mobile in the abdominal cavity, associated with long splenic pedicle and absence of peritoneal attachments.[12] Treatment options include splenectomy or splenopexy. Findings such as presence of a massive spleen or associated complications such as rupture or bleeding preclude splenopexy.[13,14] Total splenectomy is generally preferred owing to long pedicle that result in torsion and subsequent splenic infarct.[15] Epidermoid cyst of the spleen is uncommon, and in the presence of wandering spleen is exceedingly rare. To our knowledge, only two other case reports has been described of this association.[16,17]

CONCLUSION

Wandering spleen and epidermoid cysts of the spleen are two rare entities, and their coincidence is sparsely described in literature. The connection between these two is unknown, as only two other cases have been described of this association. However, the discovery of this entity warrants surgery as many are symptomatic and are prone to torsion of the spleen.

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