An epidermoid cyst of accessory spleen simulating tumors of the tail of pancreas

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ABSTRACT

An epidermoid cyst of accessory spleen, a rare condition may present as pseudocyst of pancreas and other cystic tumors of the pancreas. This case report along with the review of literature attributes some clinical features and investigative pattern to differentiate between epidermoid cyst of accessory spleen and other cystic tumor of pancreas.

Plan of exploratory laparotomy was made keeping differential diagnosis of pseudocyst of pancreas and other cystic tumors of pancreas. On exploration there was huge splenomegaly (20 × 50 × 10 cm) having soft cystic cavity that was confirmed on aspiration. There was dense adhesion around spleen and the mass in the tail of pancreas, which was continued with spleen. Splenectomy was done along with resection of mass in the tail of pancreas. The excised mass was sent for histopathological examination. Cystic fluid was sent for cytological examination.

Histologically cyst was lined with flattened cuboidal epithelium varying from single layer to 3–4 cells thick at some places. Focal squamous lining was also seen. Cyst wall was composed of fibrous connective tissue mixed with inflammatory infiltrate; focally cyst macrophages and microcalcification was also present. Splenic red and white pulp was seen. Cytological examination of fluid shows hemorrhage and few degenerating cells.

1. Case report

A 10 year old male child was admitted to our hospital with complaints of recurrent episode of pain in left hypochondrium and recurrent attacks of fever for last one year. There was no history of trauma in the past. On clinical examination, a lump (20 cm × 15 cm) was felt in left hypochondrium moving craniocaudally with respiration. Routine blood investigations were within normal limits. Serum amylase and lipase was 28 and 74 IU/l, respectively. Serum CEA (carcinoembryonic antigen) level was normal. On ultrasonography, thinned out pancreas and 16.6 × 13 × 12 cm (approx. 1360 ml) walled off collection with internal echoes in lesser sac was seen. Patient was managed initially non-operatively and on subsequent ultrasonography, almost similar findings were noted. On CT scan (Fig. 1), a large (12.9 × 12.2 cm) fluid filled cystic lesion was seen anterior to left kidney and extending lateral to pancreas and antero-medial to spleen reaching up to anterior abdominal wall. The rim of cyst was incompletely calcified. Lesion displaced stomach and bowel to right. Distal body and tail of the pancreas was not visualized separately, suggestive of pancreatic cystic tumor.

Upper gastrointestinal endoscopy was done showing large inward bulge in fundus of stomach with normal mucus covering probably due to external compression (Fig. 2).

2. Discussion

Management of a cystic lesion of the pancreas or spleen needs precise diagnosis. A cystic lesion occurring in this region ranges from several benign to malignant conditions. The occurrence of accessory spleen is not a rare condition but the occurrence of accessory splenic cyst in intrapancreatic region is extremely a rare condition. To date only 19 case has been reported [1,2]. The incidence of splenic cyst is less than 1% in splenectomized patients [3].

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These cysts are generally diagnosed incidentally due to increasing use of ultrasonography. These cysts are classified as primary or secondary on the basis of the presence or absence of the epithelial lining. Epidermoid cyst of spleen is a rare diagnosis having incidence of 10% of true nonparasitic splenic cysts [4]. Accessory spleens are located most commonly at the splenic hilum near pancreatic tail [5]. Intrapancreatic accessory spleens are rarer condition. The earliest report of accessory splenic cyst in intrapancreatic region is by Davidson et al. [6].

In this case it was very difficult to make diagnosis of epidermoid cyst of spleen or cystic tumors of pancreas on clinical or radiological examination. The differential diagnosis of cyst in this anatomical location include various benign or malignant conditions like pseudocyst of pancreas, endometrial cysts, serous and mucinous cystic tumors, intraductal papillary ductal tumors, lymphoepithelial cysts etc. It is worth mentioning that Elite et al. reported a rare case of squamous cell carcinoma in epidermoid cyst of spleen [7]. Our initial diagnosis was pseudocyst of pancreas which does not contain a lining epithelium. But here the cyst wall was lined with cuboidal epithelium. The other differential diagnoses were cystic tumors of pancreas-based on rim calcification, normal serum amylase level and no history of trauma and pancreatitis. But in this case, on exploratory laparotomy the cystic mass was occupying tail of pancreas and splenic region (Fig. 3). Similar to epidermoid cyst of spleen, the lining epithelium of epidermoid cyst of intrapancreatic accessory spleen consists of stratified squamous or cuboidal epithelium [8]. The histopathological report in this case report was epidermoid cyst as it was lined with cuboidal epithelium. Correlating clinical and radiological finding with histopathology, the important features which retrospectively may suggest epidermoid cyst were — location of cyst at the tail of pancreas, calcification of rim of the cyst and normal amylase level.

3. Conclusion

To conclude, clinical and radiological features which are suggestive of epidermoid cyst of accessory spleen are — location of the cyst, absence of any history of trauma, normal amylase level and rim calcification. Such kind of cyst should be kept into differential diagnosis of any cystic lesion in the left hypochondrium. Although this is a rare condition but it is worth mentioning in the differential diagnosis.

References