Epidermoid cyst in an intra-pancreatic accessory spleen with abnormally high cyst fluid CEA level: a case report

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Introduction

Accessory spleen is found in 15% of the general population, in which 5% of them are located near or in the pancreas. Epidermoid cyst in an intra-pancreatic accessory spleen is a rare benign lesion which is difficult to be diagnosed preoperatively. Cyst fluid analysis has been widely used to aid the diagnosis of pancreatic cysts.

Clinical history

A 28 year-old Chinese female with unremarkable past health was presented with epigastric pain. Abdominal ultrasound scan and contrast CT scan both revealed a unilocular pancreatic tail cyst measured around 1.5x1.8x1.4 cm with no internal septation nor solid area and a non-dilated pancreatic duct. CT-guided fine needle aspiration yielded small amount of light brown fluid. The cyst fluid carcinoembryonic antigen (CEA) level was 3582 ng/mL. Cytological examination showed no malignant cells. Laparoscopic distal pancreatectomy with spleen preservation was subsequently performed.

Pathological findings

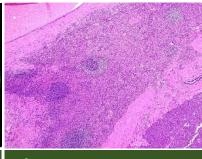
Macroscopic examination of the pancreas showed a unilocular cyst measured 1.5x1.4x1 cm and contained brown material.

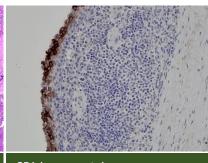
Microscopic examination showed that the cyst was lined by non-keratinizing squamous epithelium surrounded by a rim of splenic tissue, featuring white and red pulp. Immunostains for CD68 and ERG highlighted the littoral cells in the splenic tissue. The squamous epithelium was positive for p63 and CEA. The final diagnosis was epidermoid cyst in an intra-pancreatic accessory spleen (ECIAS).

Discussion

Accessory spleens are congenital and caused by incomplete fusion of mesenchymal buds in the dorsal mesogastrium during embryogenesis. Epidermoid cysts are formed from splenic mesothelial inclusion cysts with subsequent squamous metaplasia.







Contrast CT scan

H&E section

CEA immunostain

A pooled analysis of 12 studies showed that a pancreatic cyst fluid CEA cutoff value of >800 ng/mL had a sensitivity of 48 % and a specificity of 98 % for discriminating intraductal papillary mucinous neoplasms and mucinous cystic neoplasms from other non-mucinous cysts, and a low CEA level of <5 ng/mL had a sensitivity of 50% and a specificity of 95% for non-mucinous cysts, such as serous cystadenomas and pseudocysts. In our case, the cyst fluid showed high CEA level, suggesting potential pitfall in cyst fluid analysis. Studies have suggested that high CEA level in splenic epidermoid cyst is produced by the squamous epithelium, and the squamous cells are positive for CEA immunostain, which coincide with our findings.

The differential diagnoses of ECIAS histologically include lymphoepithelial cysts, dermoid cysts and retention cysts. All of them are lined by squamous epithelium. The differences are that lymphoepithelial cysts show abundant lymphocytes in the wall with germinal centre formation, dermoid cysts also contain skin appendages and columnar or respiratory epithelium, squamoid retention cysts show no lymphoid stroma and are resulted from cystic dilatation of pancreatic ducts due to obstruction.

Conclusion

ECIAS is a rare benign lesion of the pancreas which often mimics other cystic neoplasms. We should always correlate carefully with clinical and radiological findings and be cautious in interpreting cyst fluid biochemistry for preoperative diagnosis.