A carcinoembryonic antigen (CEA) value of > 1000 U/ml in pancreas cyst fluid is considered almost pathognomonic for a mucinous cystic neoplasm. A 32-year-old woman underwent abdominal computed tomography (CT) for evaluation of right upper quadrant pain, which revealed a 10-mm pancreatic tail cyst (Fig. 1a, b). The sharply demarcated cyst without septation did not communicate with the pancreatic duct. Follow-up CT, 4 months later, revealed interval growth to 16 mm. Analysis of cyst fluid obtained at endoscopic ultrasonography (EUS) revealed an elevated CEA level (1377 U/ml).

Following referral, repeat EUS revealed a 15 × 12-mm cystic mass that appeared to be extrapancreatic, abutting the pancreas and spleen. A smooth rim of tissue was noted within the cyst that had the same echodensity and pattern as the adjacent spleen (Fig. 2, Video 1). While it was unclear whether this represented retained blood secondary to the prior EUS fine-needle aspiration (FNA) or a solid component within a cystic pancreatic neoplasm, it was felt that the cyst might have been located within the spleen or an accessory spleen. However, despite the uncertainty raised by EUS as to the origin of the cyst, the CT findings and elevated CEA level all strongly suggested the presence of a cystic pancreatic neoplasm. A subsequent distal pancreatectomy and splenectomy revealed a cyst located between and densely adherent to the pancreas and spleen. Surgical pathology revealed an epidermoid cyst arising within an accessory spleen (Fig. 3a, b). Approximately 10% of benign nonparasitic splenic cysts are epidermoid cysts, which may be located within the spleen or an accessory spleen [1–3]. They are commonly associated with an elevated CA 19–9 level and the CEA level has been reported to be >1000 U/ml [4]. The pancreatic cyst differential is broad and extrapancreatic cysts may masquerade as cystic pancreatic neoplasms. Furthermore, this report highlights the fact that the CEA level is only one feature to be considered, and markedly elevated levels do not absolutely indicate a mucinous cystic neoplasm.

Epidermoid accessory splenic cyst masquerading as a pancreatic mucinous cystic neoplasm
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Fig. 3  a Epidermoid cyst within an accessory spleen. Note the epithelial cyst lining at the top, with surrounding splenic tissue inferior to it, and adjacent pancreas demonstrating chronic pancreatitis inferior to the splenic tissue (hematoxylin and eosin [H&E] stain, × 40). b A magnified view shows the stratified squamous epithelial lining of the cyst. (H&E, × 400).