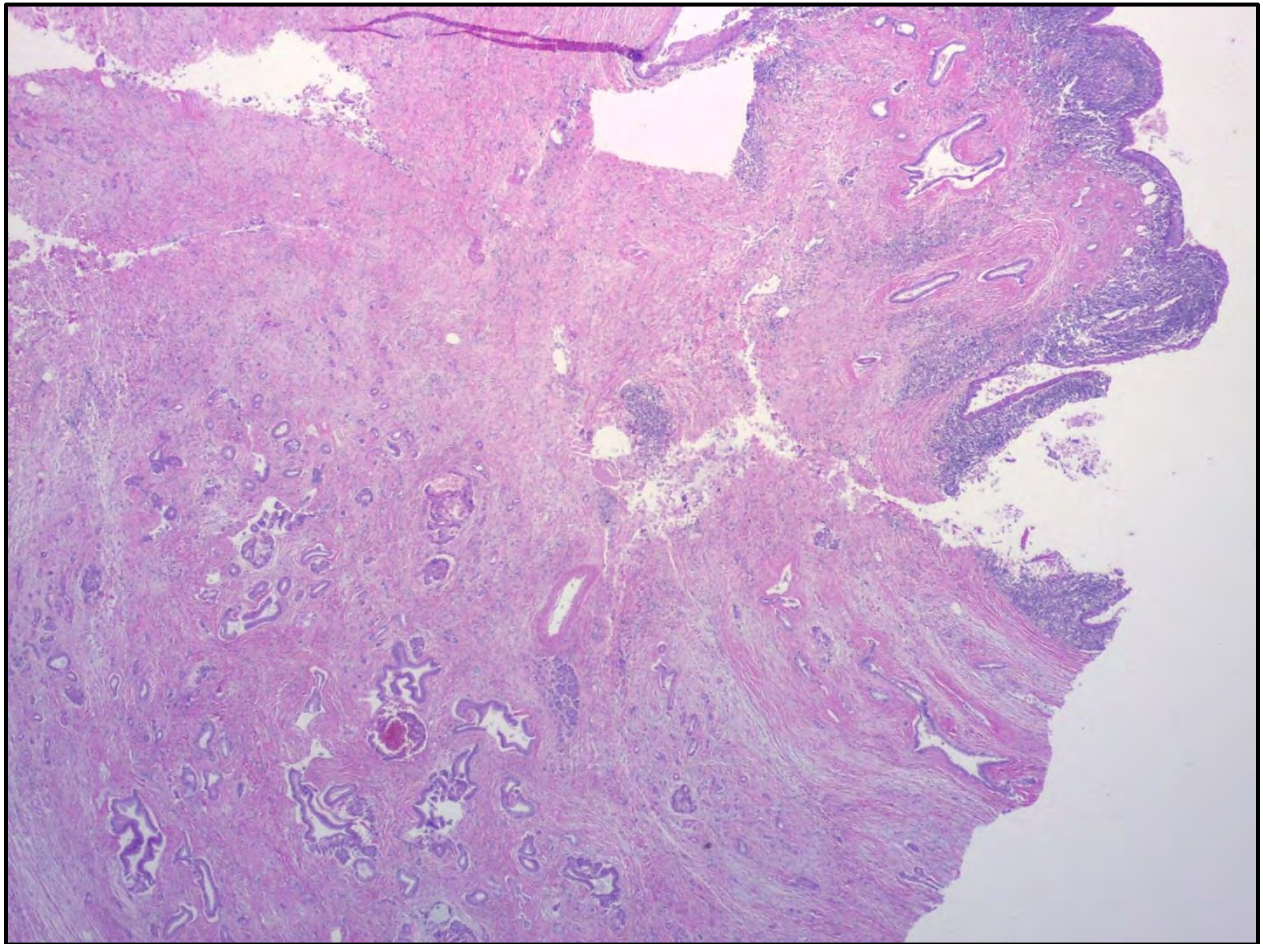
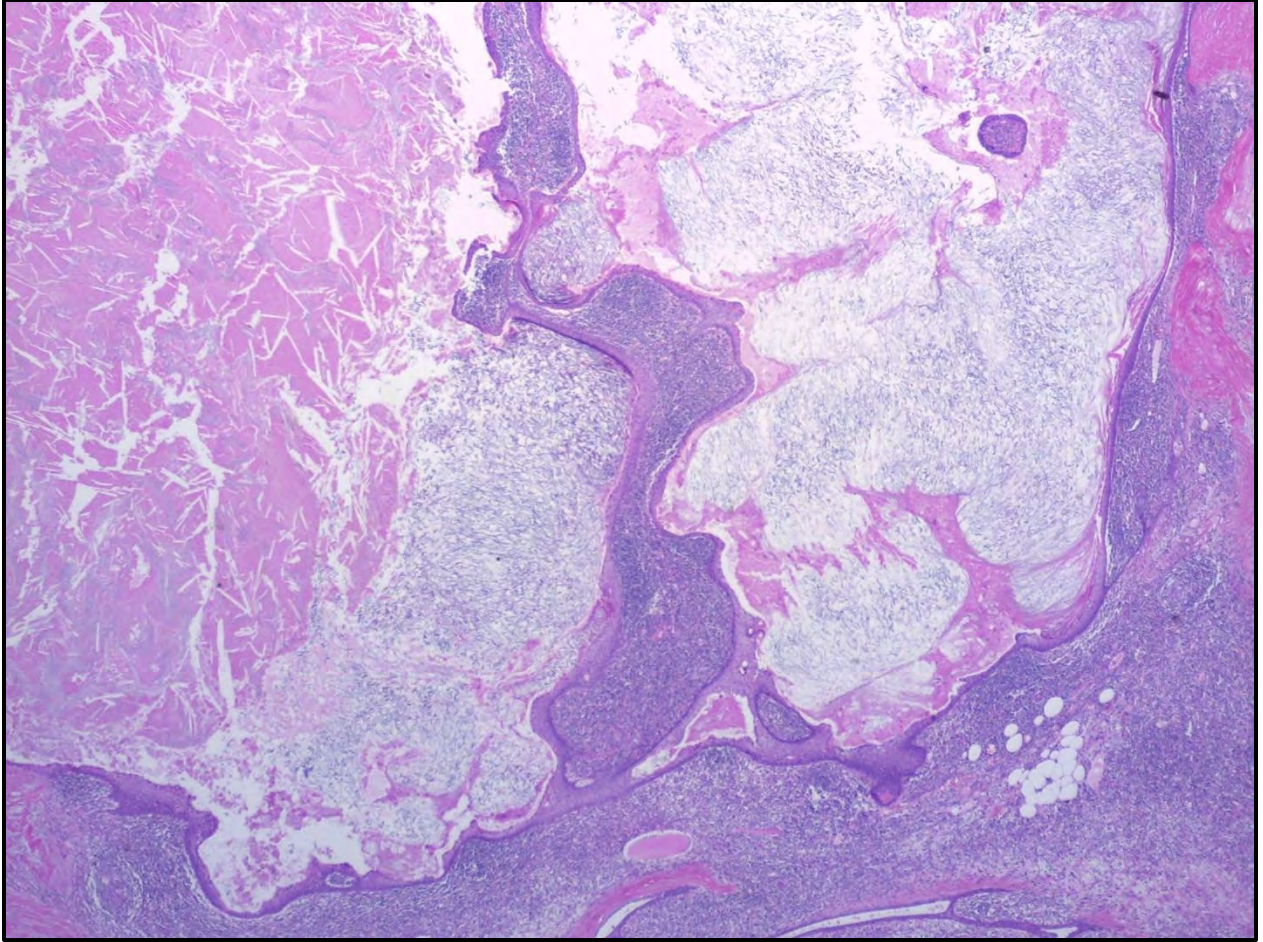
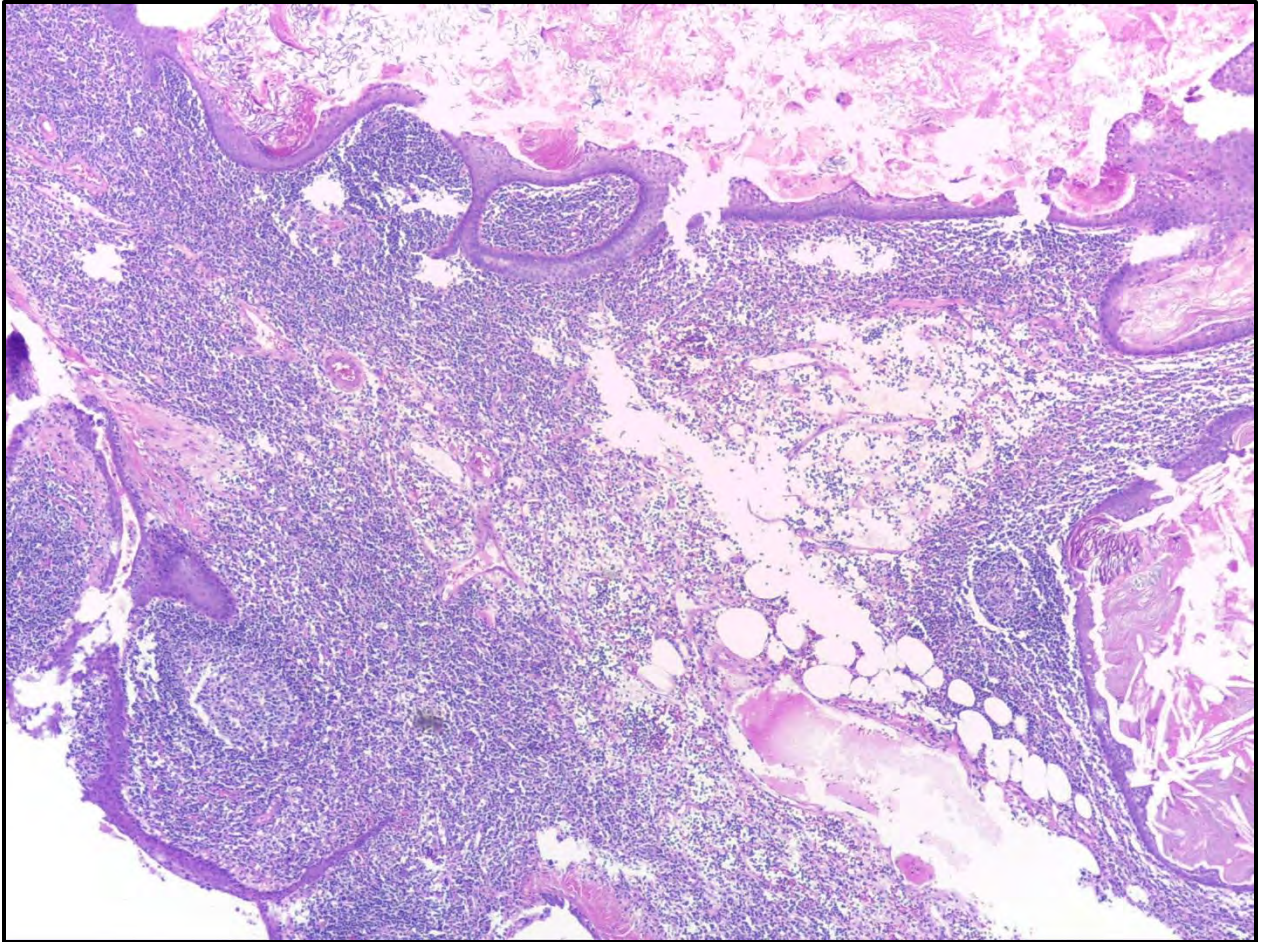
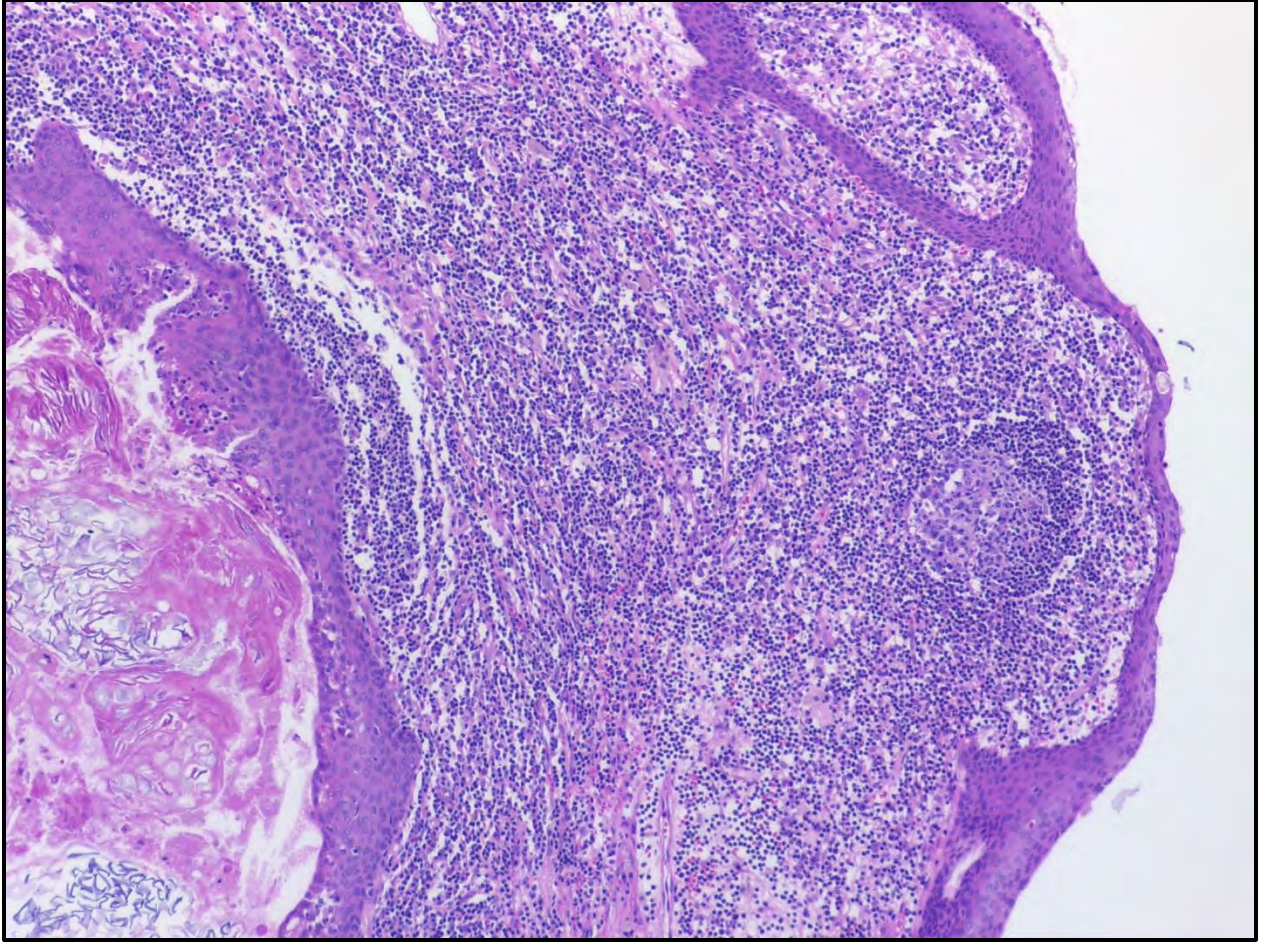


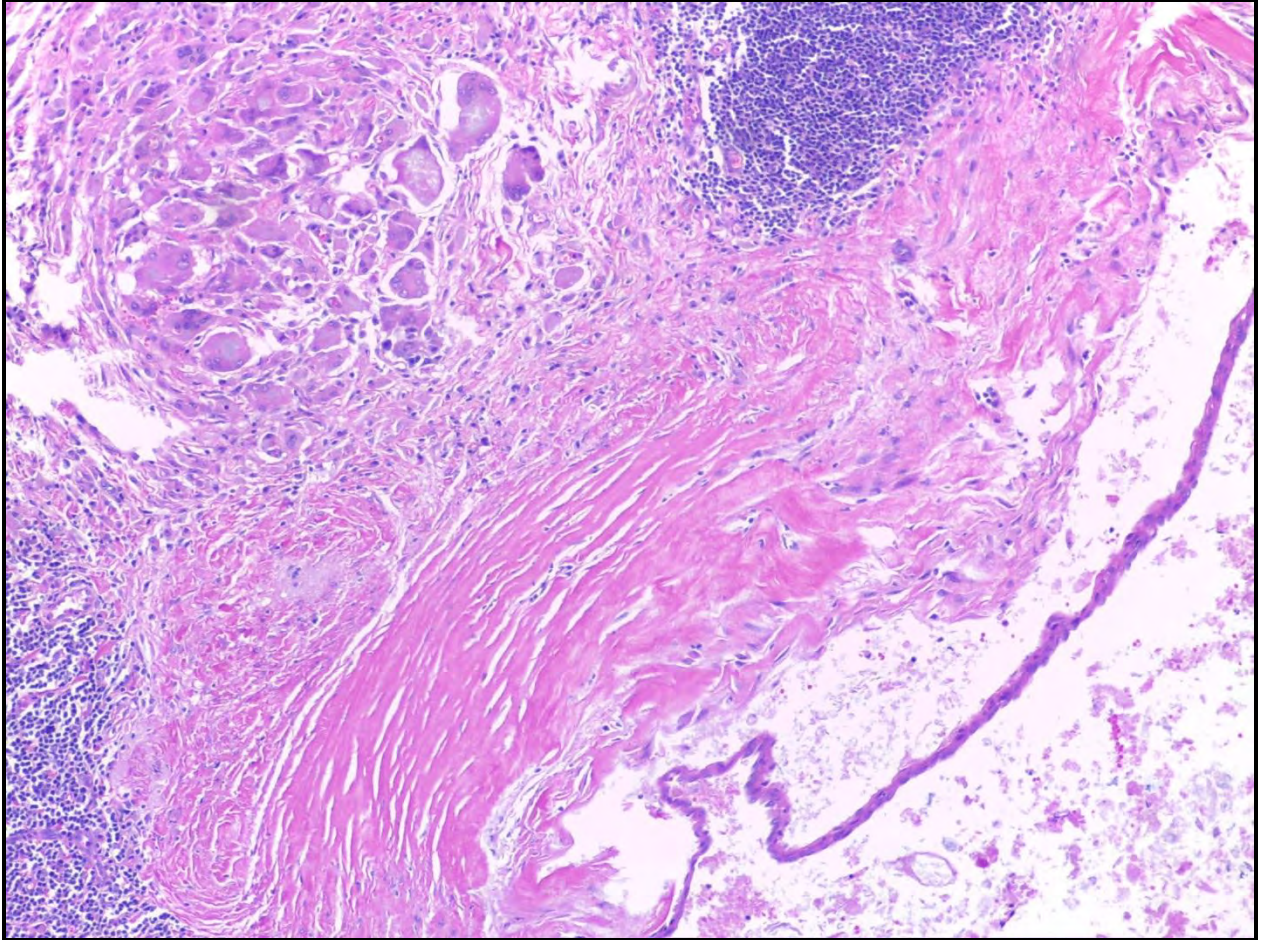
Case: 67 year old man with an incidentally discovered 6 cm cystic lesion of the pancreatic tail. Endoscopic ultrasound-guided fine needle aspiration revealed macrophages and scattered lymphocytes. Cyst fluid CEA was significantly elevated. Subsequent needle biopsy showed only fibrin and lymphoid elements with no malignant cells identified. Distal pancreatectomy and splenectomy was performed with clinical concern for an intraductal papillary mucinous neoplasm. Representative images of the resection specimen are shown below.











What is your diagnosis?

- A. Epidermoid cyst
- B. Dermoid cyst
- C. Lymphoepithelial cyst
- D. Pseudocyst

Answer and Discussion:
C. Lymphoepithelial cyst

COMMENT:

Representative micrographs show the cystic wall is lined by keratinizing squamous epithelium and underlying abundant lymphoid tissue with germinal center formation. The cyst contains abundant keratin debris with associated cholesterol clefts. Focal foreign body giant cells reactive to keratin are also seen in the surrounding fibrous tissue.

Answer A is incorrect. Epidermoid cysts in the pancreas usually occur in an intrapancreatic accessory spleen. Histologically an epidermoid cyst is characterized by a multilayered flattened or cuboidal epithelial lining, reminiscent of squamous epithelium with surrounding splenic tissue. In some cases, the cyst lining may be partly mucinous. Subepithelial lymphoid tissue is not a feature. They are located almost exclusively in the tail of the pancreas.

Answer B is incorrect. Dermoid cysts of the pancreas are also called cystic teratomas. A dermoid cyst has a cyst wall lined by keratinized squamous epithelium and underlying sebaceous glands are often seen. The cyst usually contains differentiated tissues from one or more germ cell layers, usually ectodermal. The cyst contents appear pasty, "cheesy" or "caseous", with keratinaceous and sebaceous secretions, and only rarely may be clear and serous. Mucinous epithelium, respiratory-type mucosa, sebaceous units and hair follicles are more readily identified in dermoid cysts rather than in lymphoepithelial or epidermoid cysts. Adjacent dense subepithelial lymphoid tissue that contains lymphoid follicles and germinal centers has also been reported in dermoid cysts, but other defining features of a dermoid cyst are also present.

Answer D is incorrect: Pseudocysts contain pancreatic secretions, inflammatory cells, and debris without an epithelial lining. A pseudocyst usually develops secondary to pancreatitis, trauma, ductal calculi or an adjacent obstructive neoplasm. The cyst wall consists of dense collagen with histiocytes, giant cells and granulation tissue, and no lymphoid tissue is seen. Cholesterol clefts can also be present in pseudocysts.

Answer C is correct: Pancreatic lymphoepithelial cysts can occur anywhere in the pancreas, and grossly are sharply demarcated from the surrounding tissue. The size can range from 1 cm to 17 cm. Patients often present with abdominal pain, nausea, vomiting and diarrhea, although they can also be completely asymptomatic and discovered incidentally. Interestingly, cyst fluid CEA can be elevated. Lymphoepithelial cysts are the most common squamous lined cysts in the pancreas. Malignant transformation has not been reported. Microscopically they are characterized by stratified squamous epithelium surrounded by a band of mature lymphoid tissue with intervening well-formed germinal centers. Other types of admixed epithelium, such as non-keratinizing squamous, flat cuboidal or transitional epithelium, may also be seen. Rare scattered sebaceous or mucinous cells may be seen, but adnexal structures should not be present.

References:

Adsay NV, Hasteh F, Cheng JD, Klimstra DS. Squamous-lined cysts of the pancreas: lymphoepithelial cysts, dermoid cysts (teratomas), and accessory-splenic epidermoid cysts. Semin Diagn Pathol. 2000 Feb;17(1):56-65.

Hruban RH, Pitman MB, Klimstra DS, Tumors of the Pancreas (AFIP Atlas of Tumor Pathology; 4th Series Fascicle 6). 2007, 353-364.

Raval JS, Zeh HJ, Moser AJ, Lee KK, Sanders MK, Navina S, Kuan SF, Krasinskas AM. Pancreatic lymphoepithelial cysts express CEA and can contain mucous cells: potential pitfalls in the preoperative diagnosis. *Mod Pathol*. 2010 Nov;23(11):1467-76.

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