ABSTRACT

Introduction: Enterogenous cysts of the pancreas are rare congenital foregut cysts. The natural history of these entities is still relatively unknown, as is their propensity for malignant transformation. Thus, optimal management is still undetermined.

Case Description: A 39-year-old African American woman underwent a computed tomography scan of the chest that showed an incidental cystic lesion of the pancreas. Endoscopic ultrasonography–guided fine-needle aspiration (FNA) identified an elevated carcinoembryonic antigen level, as well as benign, focally ciliated columnar cells, histiocytes, and proteinaceous material, with no evidence of malignant transformation. These findings were consistent with respiratory or bronchial versus dermoid or endometrial (müllerian) origin. A laparoscopic distal pancreatectomy was performed, and the final pathologic findings showed gastric tissue surrounded by a rim of normal pancreatic tissue, consistent with a benign enterogenous cyst. The patient recovered well.

Discussion: Enterogenous cysts that arise outside of the liver are exceedingly rare and represent both a diagnostic and therapeutic challenge. On imaging, they can be mistaken for malignant cystic lesions. Pathologic examination provides definitive identification, with FNA smears giving pathognomonic characteristics; however, FNA samples can miss the architecture of the cyst. Moreover, there is limited knowledge of the natural history of these lesions, and reports exist of other enterogenous or developmental abnormalities undergoing malignant transformation. Although it would be preferable to avoid unnecessary surgery for these rare lesions, not enough is known about their behavior. Surgery remains the standard of care and the more “conservative” management option for these cystic lesions of the pancreas.

Key Words: Enterogenous cyst, Pancreas, Laparoscopic distal pancreatectomy
Computed tomography scan of the chest showed an incidental pancreatic tail lesion (Figure 1). A magnetic resonance imaging scan was subsequently performed, showing a non-enhancing lobulated cystic lesion of the pancreatic tail suggestive of a mucinous cystic pancreatic neoplasm (Figure 2). Endoscopic ultrasonography (EUS) showed a unilocular cyst measuring 4 × 2.8 cm with cloudy beige aspirate. On pathologic analysis, the smear contained benign, focally ciliated columnar cells, histiocytes, and proteinaceous material, with no evidence of malignant transformation. These findings were consistent with respiratory or bronchial versus dermoid or endometrial (müllerian) origin. The CEA level of the aspirate was 5450 ng/mL. The serum CEA level was 0.7 ng/mL, the CA 19–9 level was 1.0 U/mL, and the amylase level was normal. After the esophagogastroduodenoscopy, the patient underwent an uneventful laparoscopic distal pancreatectomy with splenectomy. Intraoperative ultrasonography showed a cystic lesion in the pancreatic tail within the hilum of the spleen. The fine-needle aspiration (FNA) results suggested a benign pathologic course, but at the time of surgery, an enterogenous cyst was low on the differential. Not only is it a rare lesion, but also pathologic analysis suggested an endometrial origin rather than an enteric duplication cyst. The distal location and intimacy with the hilum of the spleen made enucleation challenging, requiring a splenectomy with resection of the pancreatic tail. No gross intraoperative images were taken. The pathologic findings of the specimen described an entirely benign cyst measuring 2.0 × 1.5 × 1.2 cm, composed of mucosal, submucosal, and muscular layers.

Figure 1. Computed tomography scans of incidental cystic pancreatic tail mass: axial view (A) and coronal view (B).

Figure 2. Magnetic resonance imaging scan of pancreatic tail mass (red circle).

Figure 3. Composite image of enterogenous cyst histology (H&E stain, magnification 40×). Inset A: gastric mucosa (H&E stain, magnification 100×); Inset B: respiratory epithelium (H&E stain, magnification 400×).
with a lumen lined with rare foci of gastric mucosa (Figure 3) and ciliated respiratory-type epithelium. The spleen and lymph nodes were normal. The patient had an uneventful postoperative course.

DISCUSSION

Enterogenous cysts of the foregut are defined as those “arising from the intestines” and are lined with epithelium that simulates gastric or normal intestinal tissue. Those that arise outside of the liver are exceedingly rare and represent both a diagnostic and therapeutic challenge. Recent reports have provided a tool for the classification of these lesions, but there is still no consensus on the management. Ruling out a malignant process, such as mucinous cystic neoplasms (MCNs) and intraductal papillary mucinous neoplasms (IPMNs), is imperative. Because of the rarity of these lesions, there is little understanding of their biological activity and no precedent on the best treatment. With suspicious histologic elements, such as an elevated CEA level and the presence of mucin, as well as indeterminate imaging findings, resection becomes the standard of care. Resection could, in the face of this unknown natural history, be considered the more “conservative” form of treatment, eliminating any threat of malignant transformation.

Enterogenous cysts are difficult to differentiate from other cystic lesions on imaging. Because MCNs and IPMNs have malignant potential, resection for cystic lesions is often recommended. Where enterogenous cysts differ from other cystic neoplasms is in the pathology. In our patient, the EUS aspirate identified ciliated columnar cells and histiocytes consistent with a respiratory or bronchial origin. Elevated CEA levels in the cyst are consistent with respiratory-type epithelium. This pathologic condition concurs with other case reports on enterogenous cysts. Although this pathology is pathognomonic of enterogenous cysts, the FNA does not always provide sufficient samples of the cyst wall or architecture. Definitive histologic analysis in this case included gastric tissue and histiocytes consistent with a respiratory or bronchial origin. Elevated CEA levels in the cyst are consistent with respiratory-type epithelium. This pathologic condition concurs with other case reports on enterogenous cysts.

As previously mentioned, the rarity of these pancreatic enterogenous cysts means that we do not fully understand their malignant potential. Hepatic foregut cysts, which are more common than those of the pancreas, have been noted to undergo malignant conversion. Also reported was the development of a malignancy arising from ciliated respiratory epithelium in a mature ovarian teratoma. Conversely, other reports suggested that these are developmental abnormalities and lack evidence of metaplasia. Such findings could argue that these benign developmental entities do not require surgery; however, in light of limited experience with enterogenous cysts, other models for the resection of benign processes should be considered. Surgery is recommended when there is an inability to monitor neoplastic change in benign or developmental processes. For example, choledochal cysts, lobular carcinomas in situ, or cryptogenic testes are all resected. Normal glands are resected for increased genetic risk as well (eg, thyroid in multiple endocrine neoplasia type 2B). Alternatively, there is often a mass effect with large cystic lesions, in addition to the inability to rule out the presence of a tumor, making resection the standard. The rare enterogenous cysts of the pancreas could be grouped among these benign processes for which surgery should be considered. Histologic findings on FNA may indeed indicate a benign process, but with the evidence of similar cysts undergoing dysplastic changes, a conservative approach involves surgical resection. This eliminates the concern over malignant transformation and any premalignant elements missed on FNA. Finally, resection provides a conclusive diagnosis and management for both the patient and the physician alike.

CONCLUSION

Enterogenous cysts of the pancreas are still a rare entity. Diagnostic challenges regarding this benign lesion include its similarities to malignant or premalignant cystic neoplasms that require excision for appropriate management. Because pathology is the definitive identifier of the final nature of these lesions, surgery is recommended as the conservative approach, especially in healthy patients with accessible lesions.

References:


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