Subdiaphragmatic Foregut Cyst: Case Report, Differential Diagnosis, and Review of the Literature

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ABSTRACT

Foregut-derived cysts are uncommon lesions, particularly when located below the diaphragm. When so encountered, they enter into the differential diagnosis of upper retroperitoneal masses. We report a case of such a lesion presenting as a pancreatic mass and review the current literature on this topic.

Key Words: Foregut cysts, Pancreatic neoplasms, Amylase.

INTRODUCTION

The widespread use of sophisticated imaging techniques has lead to increased discovery of pancreatic and peri-pancreatic cysts. Cystic lesions of the pancreas encompass a broad range of pathological entities and may be classified into neoplastic and non-neoplastic types. However, the classification scheme proposed by Kosmahl et al incorporates the currently described cystic lesions into 4 groups, facilitating preoperative diagnosis. The cysts are first separated into neoplastic and non-neoplastic categories, and then subdivided into those with or without an epithelial lining. The neoplastic, epithelial neoplasms are the largest group and include serous, mucinous, intraductal papillary, and solid pseudopapillary lesions. The second largest group, the non-neoplastic, epithelial lesions, consists of congenital, retention, and enterogenous cysts. Neoplastic nonepithelial neoplasms (eg, lymphangioma, and sarcomas) and non-neoplastic nonepithelial lesions (eg, pancreatitis associated pseudocyst, parasitic cysts) account for the remainder.

Differentiation between these entities is based on clinical information, associated changes in other organs, and location and character of the cyst wall. Distinction may be difficult on imaging studies alone so that serological and aspiration cytology may be used. However, even with these techniques, a specific preoperative diagnosis cannot always be determined and surgical excision is often required for both diagnosis and therapeutic management.

CASE REPORT

Ten months after the delivery of twins by Cesarean delivery, a 34-year-old woman complained of abdominal bloating. She had no additional symptoms, such as nausea, vomiting, fever, or chills. No past or present history of pancreatitis was elicited. Her past medical history included gestational diabetes, hypertension, irritable bowel syndrome, and 3 fertility related laparoscopies. On physical examination, she was noted to be obese with a non-distended, nontender abdomen. No masses were palpable. Laboratory values, including serum amylase and lipase, were normal. A CT scan and intraoperative lapa-
Endoscopic images of the abdomen and pelvis showed a 5-cm cystic mass immediately near the tail of the pancreas (Figure 1).

The patient underwent laparoscopic resection of the mass with intraoperative ultrasound-guided localization. The technique for the operation utilized one 12-mm trocar in the right upper quadrant, and three 5-mm trocars in the supraumbilical region, left upper quadrant, and lateral left subcostal margin. The left lateral lobe of the liver was reflected out of the way with a Nathanson retractor placed in the subxiphoid region. A 5-mm, 30-degree scope was used for the procedure. After the lesser sac was entered through the gastrocolic ligament with ultrasonic dissection, the mass was localized with endoscopic ultrasound. Removal was then completed with the ultrasonic dissector. The mass was removed through the slightly enlarged 12-mm port site. The patient’s postoperative course was uneventful, and she was discharged home on the second postoperative day. A 5.3x3.6x3.0-cm smooth surfaced, cystic mass was sent to the pathology laboratory. Ten milliliters of cloudy fluid was aspirated and sent for amylase analysis. The cyst lining was smooth, and the wall was 0.8-cm thick. Histological examination demonstrated a lining of mature, ciliated, pseudostratified epithelium without cartilage or mucus glands (Figure 2). The wall was composed of 2 distinct layers of smooth muscle with ganglion cells in between (Figure 3).

**DISCUSSION**

In our case, the presence of a true epithelial lining immediately discounted the possibility of a pancreatic pseudocyst. The ciliated nature of the cells suggested either bronchial type lining or, because of the absence of mucus glands and cartilage, a possible fallopian tube remnant. However, the presence of a dual layered mus-

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**Figure 1.** A, Computed tomography (CT) image of the pancreatic cyst (arrow); B and C, intraoperative laparoscopic views of the cyst in situ.

**Figure 2.** A, Low power view (4x) of the cyst demonstrating the well-formed muscular layers, H&E; B, High power view (20x) of the ciliated, pseudo-stratified epithelial lining, H&E.
cular wall with ganglion cells indicated that this was a foregut cyst.

Embryologically, foregut cysts arise from pinched off buds of the endodermal foregut.7–11 Cysts that form and remain in the mediastinum almost always show either bronchogenic or esophageal differentiation. Subdiaphragmatic cysts arise from gastric or small intestinal epithelium. In all of these instances, the cysts most often lie close to and recapitulate the structure of the native organ. Sometimes buds pinched off the supradiaphragmatic foregut migrate into the abdominal cavity via the pericardio-peritoneal canal before fusion of the diaphragm in the sixth week of embryogenesis.7–11 They are commonly located to the left of the midline, within a triangle behind the stomach as defined by the midline, splenic vein, and spleen/diaphragm (Figure 4),12 as in our case. Pancreatic and peri-pancreatic foregut cysts are rare lesions, with about 50 cases reported in the English literature to date.12

Clinically, patients with subdiaphragmatic foregut cysts may present with abdominal or flank pain, with or without accompanying nausea and vomiting. More frequently, however, these lesions are incidentally identified, occurring in the absence of laboratory aberrations. Radiologically, these cysts have been described as unilocular, fluid-filled masses with rare calcifications of the wall, ranging in size from 0.3 cm up to 18.9 cm with a mean of 6.8 cm.12–14 Their malignant potential is virtually nonexistent, and if a precise preoperative diagnosis can be made, these lesions can be left alone.

**Figure 3.** High power view (20x) of ganglion cells and nerve bundles identified within the muscular layers of the cyst, H&E.

**Figure 4.** Triangle of subdiaphragmatic foregut cysts as indicated by the midline, splenic vein, and diaphragm/spleen. From Gray’s Anatomy (1918).
However, surgical resection is performed because of difficulties in definitive exclusion of a cystic neoplasm.

The histologic appearance of these cystic lesions follows their embryological derivations. Therefore, esophageal cysts are lined by squamous mucosa, with a wall composed of well-defined muscular layers and neural tissue. Bronchogenic cysts are lined by a ciliated respiratory epithelium, with interspersed glandular cells and areas of cartilage. Enterogenic cysts are often composed of gastric, duodenal, or pancreatic epithelium, with or without an associated muscular wall. Foregut cysts of an indeterminate nature may contain one or more components from any of the above-listed cysts, thereby making a specific diagnosis difficult. Our case demonstrated a ciliated, columnar/pseudostratified lining epithelium, suggesting a bronchial origin, with 2 well-formed muscular layers and associated ganglion cells, suggesting esophageal or enteric differentiation. Other authors\(^{12}\) have referred to this combination as being of the esophageal type. We suggest that the label “indeterminate” would be more appropriate because of the rather hybrid combination of components.

Unexpectedly, our cyst contained 16,161 U/L of amylase, which is difficult to explain in an epithelial-lined cyst. We sought hard, but did not find, pancreatic elements in the cyst wall. The cyst was separate from the pancreas, seemingly too far for amylase to diffuse into it. In the event that preoperative fluid analysis had been performed, a diagnosis of pancreatic pseudocyst would likely have been made, leading to a markedly different therapeutic approach. Other authors have noted that amylase is occasionally present within subdiaphragmatic foregut cysts,\(^ {15-18}\) which indicates that amylase analysis cannot wholly distinguish a pseudocyst from a cystic neoplasm. It would be worth investigating whether measurement of other pancreatic enzymes would be more accurate in this regard.

**CONCLUSION**

Foregut cysts are often easy to identify and explain, because they are often adjacent to, and contain the same elements as, the native organ of derivation. However, as illustrated in this case, these congenital cysts can occur in aberrant locations and may contain a mixture of histologic elements. We therefore propose the term indeterminate cysts to describe these cases. Additionally, we propose that additional investigation of amylase levels and perhaps other enzymes as well (lipase, trypsin, and others) may aid in the differentiation and preoperative diagnosis of foregut cysts from pseudocysts.

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