

## CASE REPORT

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# A Foregut Cystic Neoplasm with Diagnostic and Therapeutic Similarities to Mucinous Cystic Neoplasms of the Pancreas

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### ABSTRACT

**Context** Greater utilization of cross-sectional abdominal imaging has increased the diagnostic frequency of cystic neoplasms of the pancreas. The “International Consensus Guidelines 2012 for the Management of IPMN and MCN of the Pancreas” illustrates a diagnostic and therapeutic algorithm for these lesions based on current knowledge. **Case report** We present a case of a 49-year-old woman with two years of intermittent epigastric pain found to have an 8.5 cm head of the pancreas mass on CT. Evaluation was consistent with a mucinous cystic neoplasm for which she underwent an uneventful pancreaticoduodenectomy. Histology revealed a bronchogenic cyst of the head of the pancreas. **Discussion** Bronchogenic cysts are congenital anomalies of the ventral foregut that can migrate into the abdomen prior to fusion of the diaphragm. They can easily be misdiagnosed for other benign and malignant retroperitoneal lesions. Similarly to mucinous cystic neoplasms, bronchogenic cysts have been reported to undergo malignant transformation. They can also become infected and hemorrhage. Therefore, resection should be performed in appropriate risk candidates. It is possible, with increased use of high resolution cross-sectional imaging, that these lesions may be identified with greater frequency in the abdomen and confused with other pancreatic neoplasms. The presence of ciliated respiratory epithelium and cartilage on pathology provides for definitive diagnosis.

### INTRODUCTION

Greater utilization of cross-sectional abdominal imaging has increased the frequency of cystic neoplasms of the pancreas being brought to the attention of physicians [1]. Zhang *et al.* in 2002 discovered pancreatic cystic lesions in 19.9% of 1,444 patients studied by MRI, nearly 52% of which were not simple in nature [2]. A study of 2,832 patients undergoing CT demonstrated pancreatic cystic lesions in 2.6% [3]. Pancreatic cystic neoplasms of concern are essentially divided

between intraductal papillary mucinous neoplasms and mucinous cystic neoplasms, as serous cystic neoplasms and pseudocysts are benign [4]. The latter two lesions can also be more conclusively differentiated by non-invasive imaging and clinical history of pancreatitis or alcohol abuse, respectively. The “International Consensus Guidelines 2012 for the Management of IPMN and MCN of the Pancreas” provides a diagnostic and therapeutic algorithm based on current knowledge [5].

Nevertheless, obtaining a definitive diagnosis preoperatively may be challenging. Observing loculations, a capsule, communication with the pancreatic duct, cyst morphology, calcifications or mural nodules on cross-sectional imaging can help inform the differential diagnosis. However, typical features are often absent and this explains the varying diagnostic accuracy reported in the literature [6]. Even in an era of high quality cross-sectional imaging, age, gender, clinical history

Received January 16<sup>th</sup>, 2013 – Accepted May 7<sup>th</sup>, 2013

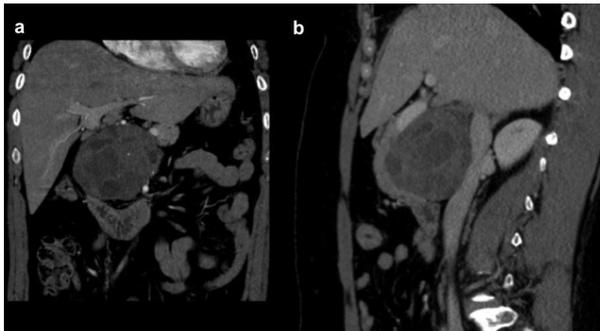
**Keywords** Bronchogenic Cyst; Neoplasms, Cystic, Mucinous, and Serous; Pancreatic Neoplasms

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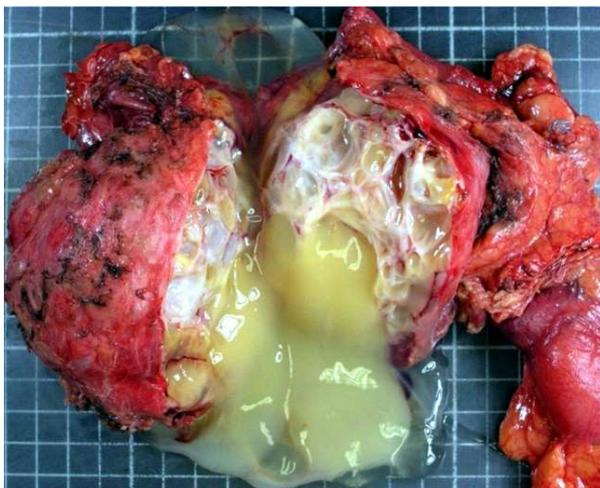
**Figure 1.** Coronal (a.) and sagittal (b.) views of an 8.5 cm head of the pancreas mass.

and the location of the cystic lesion frequently drive clinical decision making.

We describe a rare congenital lesion typically confined to the chest, but given the atypical location, it was difficult to discriminate from a mucinous cystic neoplasm based on clinical presentation and radiographic features. To our knowledge, this is the first report of this lesion being identified within the true pancreatic parenchyma.

#### CASE REPORT

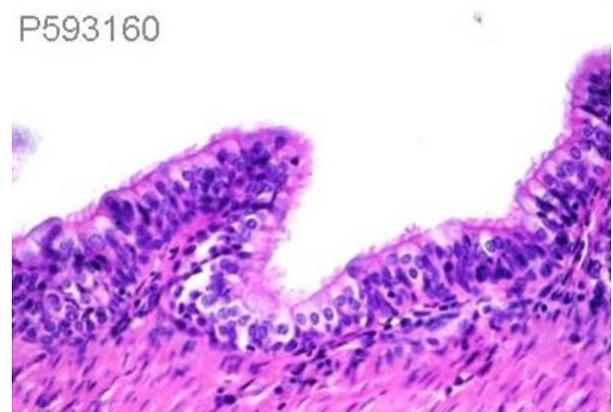
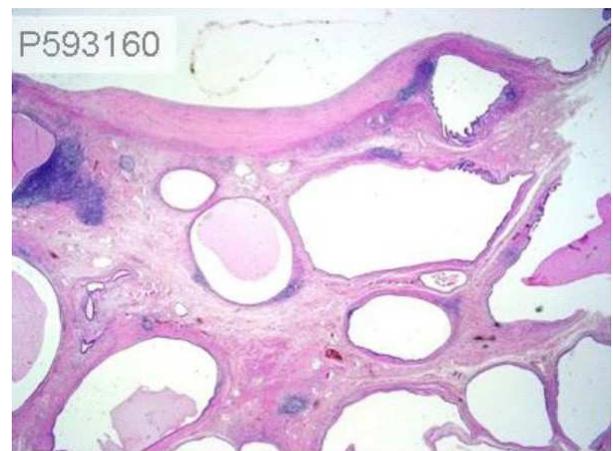
A 49-year-old woman presented with two years of intermittent epigastric pain. The episodes typically lasted a few days and were associated with nausea and anorexia. Her past medical, surgical, social and family history were non-contributory. Physical exam was only remarkable for a body mass index of 33 kg/m<sup>2</sup>, and common laboratory values were within normal range. A triple-phase CT demonstrated an 8.5 cm head of the pancreas mass with both solid and cystic components displacing the duodenum and pancreas anteriorly, and the vascular structures medially (Figure 1). There was no biliary or pancreatic ductal dilatation, and chest



**Figure 2.** Gross pancreaticoduodenectomy specimen demonstrating a well encapsulated mass comprised of multiple cysts separated by thick septae containing dense mucous. The cyst emerged from the uncinate process.

and liver imaging were unremarkable. The patient was subsequently referred to our hospital for further evaluation, at which time CA 19-9 and carcinoembryonic antigen levels were found to be normal. Endoscopic ultrasonography demonstrated a head of the pancreas mass with both solid and cystic components, thickened cyst walls, a fat plane between the vascular structures, and no suspicious lymphadenopathy. Fine needle aspiration revealed mucin, and benign cytology.

A mucinous cystic neoplasm was thus suspected. The decision was made to proceed with pancreaticoduodenectomy after discussion at multidisciplinary tumor board because of her recurrent symptoms, young age, excellent functional status, and the malignant potential of the lesion [5]. Her peri-operative course and recovery were unremarkable. Pathology demonstrated a multi-loculated cystic mass measuring 12x9x7 cm filled with dense mucous (Figure 2). Histology revealed simple and pseudostratified, ciliated, mucus secreting cells and hyaline cartilage, supporting the final diagnosis of a bronchogenic cyst in the head of the pancreas (Figure 3). There was no cellular atypia noted.



**Figure 3.** H&E staining demonstrates multiple microcysts with intervening cartilaginous elements (top) and ciliated respiratory epithelium (bottom).

## DISCUSSION

Bronchogenic cysts are rare occurrences in the abdomen, without a clear predisposition based on sex, ethnicity or age [7, 8]. As such they are easily misdiagnosed for other benign and malignant retroperitoneal lesions such as adrenal tumors, pseudomixomas, or lymphangiomas. They are congenital anomalies resulting from abnormal budding of the tracheobronchial tree from the ventral foregut during early gestation and are typically found in the posterior mediastinum. However, caudal migration prior to fusion of the diaphragm through the pericardioperitoneal channel allows for entry into the abdominal cavity. Association with the pancreas accounts for less than ten reported cases in the English literature, and all have been found on the left side of the abdomen in a triangle defined by the midline, splenic vein and diaphragm [7, 8, 9, 10, 11, 12]. This location is where 82% of all reported sub-diaphragmatic bronchogenic cysts were described in one series, with only 16% (n=6) of cysts being found on the right side of the abdomen [8].

The current case is atypical in that the lesion was associated with the posterior surface of the pancreas to the right of the superior mesenteric artery, with the portal vein and duodenum stretched over its anterior surface. This is also an uncommon location (5%) for mucinous cystic neoplasms, but there is not a reliable way to differentiate these two lesions on imaging or cytology [4]. In fact, high-resolution cross sectional imaging is often inconclusive in discriminating pancreatic cystic neoplasms with atypical features. MRI is superior to CT, and has the added advantage of simultaneously studying the lesion, the ductal system and the pancreatic parenchyma in a single study [13]. Even if a slight hyperintensity or isointensity to skeletal muscle on T1-weighted images reported for thoracic bronchogenic cysts would have been observed (MRI was not performed), preoperative diagnosis would have been difficult given the atypical sub-diaphragmatic location [14].

Mucinous cystic neoplasms have a propensity for in-situ and invasive malignancy with increasing size, and resection is recommended for surgically fit patients [5, 15]. Similarly, bronchogenic cysts have been reported to undergo malignant transformation [16, 17]. Further indications for resection include mass effect and prevention of hemorrhagic and infectious complications [9, 17].

In conclusion, we report a rare case of a sub-diaphragmatic bronchogenic cyst that because of the uncommon association with the pancreatic gland, the peculiar location (right side of the abdomen) and imaging characteristics similar to

those of a mucinous cystic neoplasm, made preoperative diagnosis problematic. The prevalence of bronchogenic cysts within the mediastinum and abdomen has not been defined. It is possible, with increased use of high resolution cross-sectional imaging, that these lesions may be identified with greater frequency in the abdomen and confused with other pancreatic neoplasms. The presence of ciliated respiratory epithelium and cartilage on pathology provide for definitive diagnosis.

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**Conflict of interest** The authors do not have any conflicts of interest, including financial, consultant, institutional and other relationships that might lead to bias in the preparation of this manuscript

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