Pancreatic Pseudocyst as a Cause for Secondary Achalasia

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Secondary achalasia is commonly due to cancer. Benign causes are rare and an association with pancreatitis has never been made. Our patient presented with clinical, manometric, endoscopic, and radiographic features of achalasia that occurred in conjunction with a pancreatic pseudocyst extending into the mediastinum. Documentation of this unique relationship was made on two occasions after episodes of complicated pancreatitis and was confirmed by restoration of normal esophageal function after drainage of the pseudocyst. Onset of dysphagia occurring in association with pancreatitis suggests the presence of a pseudocyst and secondary achalasia.

Secondary achalasia has commonly been associated with cancer-related causes (1-20) and benign etiologies are rare (21,22). We report a case of secondary achalasia resulting from a pancreatic pseudocyst with mediastinal extension. To our knowledge, this relationship has not been previously described.

Case Report

A 59-yr-old man was admitted to the hospital because of abrupt onset of dysphagia for solid and liquid foods, which began 6 wk before admission. Regurgitation of undigested food, anorexia, and a 17-kg wt loss were noted during this interval. Eight weeks before admission the patient complained of severe epigastric pain following an alcohol binge that resolved without medical attention. The past medical history was remarkable for chronic alcohol abuse.

Physical examination, stool Hemoccult, and laboratory test results including complete blood count, SMA20, amylase, lipase, urinalysis, and chest x-ray were normal. A barium esophagram showed a dilated esophagus with a smooth, tapered, conical narrowing of the distal segment (Figure 1).

Esophageal manometry was performed using a 4-lumen polyvinyl catheter (luminal diameter, 0.8 mm) perfused at 0.5 ml/min by a low-compliance pneumohydraulic capillary system driven by a nitrogen pressure head of 16 psi (Arndorfer Medical Specialists, Greendale, Wis.). The pressure rise rate was >300 mmHg/s. Radially oriented side ports were spaced 5 cm apart. The catheter was attached to a Beckman 9877 coupler on a R611 physiograph recorder (Beckman Instruments Inc., Schiller Park, Ill.). Chart speed was 2.5 mm/s and the station pull-through method was used. Manometry showed aperistalsis and failure of the lower esophageal sphincter to relax after deglutition (Figure 2). Intraesophageal resting pressure was not elevated and the lower esophageal sphincter pressure (17 mmHg) was normal. Esophagogastroduodenoscopy showed a dilated esophagus without peristalsis. The endoscope could be passed from esophagus to stomach without resistance, and the mucosae of these structures were normal.

One day after endoscopy the patient noted resolution of his symptoms and ate without dysphagia or regurgitation. Computed axial tomography performed after esophagogastroduodenoscopy showed a 5-cm pancreatic pseudocyst impinging on the cardial region of the lesser curvature of the stomach and small punctate calcifications consistent with chronic pancreatitis. Esophageal manometry performed 4 wk later showed return of peristalsis, normal relaxation of the lower esophageal sphincter with deglutition, and a resting pressure of 10 mmHg. A repeat esophagram was normal and serial computed axial tomography scans showed progressive resolution of the pseudocyst.

The patient remained asymptomatic for 6 mo before presenting with abrupt onset of dysphagia for solid and liquid foods. Serum amylase was 212 U/L (range, 27-116 U/L). A barium esophagram showed a smooth, tapered, distal narrowing identical to the first x-ray. A computed axial tomography scan showed a 3.5-cm pancreatic pseudocyst that extended into the mediastinum, impinged on the heart, and extrinsically compressed the distal esophagus (Figures 3A and 3B). Esophageal manometry showed simultaneous and repetitive contractions, aperistalsis, and failure of the lower esophageal sphincter to relax following deglutition. A pericardial friction rub was auscultated and
Figure 1. Barium esophagram (oblique projection) shows distal narrowing suggestive of achalasia.

echocardiography demonstrated a moderate-sized anterior and posterior pericardial effusion without evidence of tamponade. Internal drainage of the pseudocyst was achieved by cystogastrostomy. Postoperatively, dysphagia and the pericardial effusion resolved. One month after surgery esophageal manometry [Figure 4], an echocardiogram, a computed axial tomography scan, and a barium esophagram were normal and the patient was asymptomatic.

Discussion

The features of primary achalasia are well known (23). Secondary achalasia is manometrically identical to the primary disorder but an underlying disease is responsible for the motor abnormality. The majority of cases are due to malignancy, and gastric adenocarcinoma is the most common cause. Overall, 2%-4% of achalasia cases are due to secondary causes and benign etiologies are extremely rare (1,24). Excluding Chagas' disease, only an isolated case of amyloidosis and 1 patient with truncal vagotomy and antrectomy have met manometric criteria for diagnosis of achalasia (21,22).

The pathophysiology for secondary achalasia has not been precisely defined. Purported mechanisms include tumor infiltration with neural damage (25,26), a paraneoplastic effect (27), esophageal obstruction causing neuromuscular contractile damage (28,29), effects of postsurgical vagotomy (22), or

Figure 2. Esophageal manometry illustrates aperistalsis and simultaneous contractions in the esophageal body (A, B, C) and failure of the lower esophageal sphincter to relax (D). WS, wet swallow. Scale in mmHg.
dorsal ganglion damage (30, 31). Vagal stimulation is responsible for inhibition of the intrinsic tonic contractile state of the lower esophageal sphincter. Interruption of vagal influence by any one of these possible mechanisms could result in secondary achalasia. The pathophysiologic events causing achalasia in the case of a pseudocyst are unknown, but vagal neuropathy from compression or a nonspecific response to partial esophageal obstruction is possible. In our patient there was rapid clinical improvement in esophageal function after upper endoscopy and surgery, and this observation supports the latter mechanism. Although physiologic events are unclear, it is likely that mediastinal extension of the pseudocyst is required for the achalasia-like syndrome to occur. The prompt resolution of symptoms that followed esophagogastroduodenoscopy probably occurred because the pseudocyst repositioned itself from thorax to abdomen due to mechanical effects of passing the endoscope or from diaphragmatic motion from procedure-related gagging or retching.

Less than 60 cases of mediastinal pseudocysts have been reported in the literature (32-50) and alcohol was the cause for pancreatitis in half of these. The diagnosis was made preoperatively in ~50% of cases. Pseudocysts extended into the mediastinum through the esophageal or aortic hiatus, foramen of Morgagni, or a diaphragmatic rent. In one series, radiographic evidence of compression or displacement of the esophagus was present in 61% of cases and in 10% of individuals dysphagia was the presenting complaint (44). Esophageal motility studies were not performed.

In addition to benign secondary achalasia, our patient had an associated pericardial effusion that may have been related to the pseudocyst. The association of a pseudocyst and a pericardial effusion has been observed on two occasions (43, 51). Both patients are symptomatic and required pericardiocentesis or partial pericardiectomy. In each case, the
pericardial fluid contained markedly elevated levels of amylase. In our case, it is possible that contact between the pseudocyst and the pericardium had an irritative effect and caused fluid accumulation in the pericardial sac. However, the pathophysiology for this problem is unknown and pericardial effusions in patients with uncomplicated pancreatitis have also been observed.

Our patient’s initial presentation was consistent with achalasia but his age, degree of weight loss, and short duration of symptoms suggested a secondary cause (28). The computed axial tomography scan was performed to detect neoplastic involvement of the lower esophageal sphincter that was not apparent by other means. Identification of the pancreatic pseudocyst in proximity to the esophagus suggested the possibility of benign, secondary achalasia. Treatment for this disorder is always directed toward the underlying cause (1,14,17,18,21,22,29). Initially, no specific therapy was offered because the patient’s symptoms resolved with spontaneous reduction in size of the pseudocyst. However, cystogastrostomy was eventually required for decompression of the pseudocyst. Resolution of esophageal symptoms should follow successful drainage of the pseudocyst.

References


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