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Got Milk? An Unusual Cause of Abdominal Pain

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A 43 year old woman with a history of abdominal trauma from a motor vehicle accident three years prior to admission presented with increasing severe abdominal pain located in the epigastrium and left upper quadrant with radiation to the back, significant nausea, and intermittent vomiting. The pain was exacerbated by both eating and drinking. On physical examination she was afebrile with a soft abdomen that was tender to minimal palpation in the epigastric and left upper quadrant regions. Laboratory data was notable for normal amylase, lipase, liver function tests, white blood cell count, and hematocrit. The pain improved minimally with NPO status and intravenous fluids. Initial magnetic resonance imaging (MRI) of the pancreas before admission demonstrated pancreatitis involving the neck and body of the pancreas with peripancreatic fluid. A follow-up MRI was performed (Figures A and B). Endoscopic ultrasound subsequently confirmed the presence of multiloculated cysts adjacent to the neck-body of the pancreas with the largest pocket measuring 2.5 by 1.6 cm and normal-appearing adjacent pancreatic parenchyma and pancreatic duct (Figure C). Endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) yielded this fluid (Figure D). What is the diagnosis?

EUS-FNA revealed milky fluid with carcinoembryonic antigen 1.8 ng/mL, amylase 307 U/L, lipase 382 U/L, and triglyceride 2972 mg/dL. During subtotal pancreatectomy, a cystic mass was identified involving the pancreatic body on the left with a normal tail to the right in the image (Figure E). Heavy cream delivered via a nasogastric tube in the duodenum produced milky fluid seen leaking from lymphatics and the cyst cavity. Most commonly presenting in children, lymphangiomas are rare benign neoplasms that arise from obstructed lymphatic vessels, which progressively dilate cystically due to detachment of lymphatic tissue during embryologic development.¹ First described by Koch in 1913, pancreatic lymphangiomas are exceedingly rare, representing less than 1% of all lymphangiomas and 0.2% of all pancreatic lesions.² Histology demonstrates fat with dilated thin walled lymphatic spaces lined by endothelium without atypia (Figure F) and pancreas infiltrated by these dilated lymphatic spaces (Figure G). Abdominal trauma, inflammation, surgery, and radiation therapy have all been associated with lymphatic channel obstruction leading to vessel dilation and potentially lymphangioma. Usually asymptomatic, patients with pancreatic lymphangioma may present with abdominal pain, nausea, weight loss, palpable

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abdominal mass, hemorrhage, hydronephrosis, or infection. CT and MRI typically demonstrate a multicystic, well-defined, encapsulated lesion with thin septa and a fatty component.³ MRI of our patient demonstrated septated, well-defined peripancreatic fluid with lymphangioma infiltrating pancreatic parenchyma in the neck and body. EUS-FNA can help diagnose lymphangiomas, and while the classic milky white fluid is not always encountered, this appearance combined with an elevated triglyceride level is diagnostic of lymphangioma.¹ After EUS-FNA, our patient reported mild improvement in her abdominal pain although she continued with poor nutrition due to postprandial pain. While complete surgical resection offers definitive therapy for pancreatic lymphangiomas, it is reserved for symptomatic patients, and recurrence can occur with incomplete excision. Following resection, our patient resumed a normal diet without abdominal pain.

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Figure A.

Figure B.

Figure C.

Figure D.

Figure E.

Figure F.

Figure G.