Case Presentation

A 48-year-old man with a history of recently diagnosed acute myelogenous leukemia (AML) presented to the emergency room with a one-month history of abdominal pain and bloating. He also reported a twenty-six pound weight loss. Review of systems was negative for fever, chills, or blood in his stools. Physical exam demonstrated jaundice but was otherwise unremarkable. Laboratory studies revealed elevated conjugated bilirubin, total bilirubin, alkaline phosphatase, gamma-glutamyl transpeptidase (GGT), aspartate aminotransferase (AST), and alanine transaminase (ALT). Amylase, and lipase were normal. The patient then underwent an abdominal ultrasound followed by CT scan (Fig).

Figure. Transverse gray scale ultrasound image (A) shows an ill-defined hypoechoic mass centered in the pancreatic head (arrow). Axial CT image with oral and IV contrast at the level of the pancreas (B) demonstrates a large, lobulated, heterogeneous mass centered in the pancreatic head (arrow). The mass compresses the adjacent duodenum. Also seen is mesenteric and retroperitoneal adenopathy with a nodal mass in the left para-aortic space. Axial CT image more cephalad within the upper abdomen (C) reveals splenomegaly with periportal adenopathy.
Key imaging finding

Pancreatic mass with adenopathy and splenomegaly in a patient with AML.

Differential diagnoses

- Granulocyte sarcoma
- Primary Adenocarcinoma
- Pancreatitis
- Metastatic disease
- Neuroendocrine neoplasm

Discussion

Pancreatic masses are relatively common in abdominal imaging. The most common etiologies are pancreatic adenocarcinoma and pancreatitis, which may simulate a discrete pancreatic mass when focal. Metastatic disease and neuroendocrine tumors are less common. Additional rare masses, such as granulocyte sarcoma, should be considered in unique clinical settings. Having a basic understanding of the imaging manifestations of these processes will assist in suggesting the correct diagnosis and guiding management and appropriate therapy.

Granulocyte sarcoma.

Granulocyte sarcoma (GS) is also known as myeloid sarcoma, chloroma, and myeloblastoma. These tumors are composed of extramedullary immature myeloid cells. This was initially described by Burns in 1811 with King in 1853 using the term chloroma (because of the associated gross pathologic green appearance) and Rappaport using the term granulocytic sarcoma in 1966. Dock made the association of leukemia with GS. The lesions have been described throughout the body.

GS is most commonly seen with acute myelogenous leukemia (AML) and less commonly with chronic myelogenous leukemia (CML) or other myeloproliferative disorders. These lesions typically occur as a manifestation of leukemia or may occur as a sign of AML in a non-leukemic patient and are associated with a blast crisis or leukemic transformation. Autopsy series suggest a prevalence of 8% in patients with the diagnosis of AML. A cytogenetic irregularity has been identified in some leukemic patients with granulocyte sarcoma, and it involves the translocation of chromosomes 8 and 21. In this translocation leukemia, granulocyte sarcoma is seen in 18 to 24% of cases.

On CT, granulocyte sarcoma presents as a solid, contrast enhancing lesion with associated adenopathy and splenomegaly. These tumors are usually pliable and may result in biliary stasis. In a patient with recently diagnosed acute myelogenous leukemia and imaging features discussed above, granulocyte sarcoma is the leading diagnosis. The patient’s described symptoms of abdominal pain and bloating are likely secondary to duodenal extrinsic compression.

The patient underwent a CT guided biopsy and the tissue samples demonstrated an unusual population of lymphoreticular cell elements and patchy atypical lymphoid infiltrates. In order to fully characterize these neoplastic cells, specific immunohistochemical techniques (such as myeloperoxidase and myeloid associated antigens) are required to differentiate granulocyte sarcoma from malignant lymphoma, which may otherwise appear similar on pathologic evaluation.

Adenocarcinoma.

Adenocarcinoma is the most common primary neoplasm of the pancreas and most commonly involves the pancreatic head. This entity on CT presents as a solid mass. Adenocarcinoma usually produces significant mass effect, resulting in dilatation of the common bile duct and pancreatic duct (the double duct sign). It is not the favored diagnosis in this case given the presence of splenomegaly and absence of biliary ductal dilatation.

Pancreatitis.

Pancreatitis has may occasionally be indistinguishable from a discrete pancreatic mass. Focal pancreatitis involving the head, "groove pancreatitis," may appear mass-like and is usually interposed between the duodenum and the pancreatic head. Acute inflammation of the pancreas, diffuse or focal, classically demonstrates peri-pancreatic fat
stranding. Patients will also have elevated amylase and lipase, which were normal in this patient. Chronic pancreatitis may have normal laboratory values but often demonstrates parenchymal atrophy, calcifications, and ductal enlargement.\(^8\)

**Metastatic disease.**

Primary neoplasms prone to pancreatic metastases include lung, breast, gastrointestinal tract, renal cell, melanoma, lymphoma, and osteosarcoma.\(^9\) Metastatic involvement is always a consideration when a solid lesion is present in a visceral organ. Typically, metastatic disease involves multiple organs with a variable appearance. Solitary pancreatic metastatic involvement may be seen with metastatic disease from renal cell, lymphoma, and melanoma.\(^9\)

**Neuroendocrine neoplasms.**

Neuroendocrine neoplasms are pancreatic islet cell tumors (ICT) which produce and secrete hormones.\(^10\) Functioning, syndromic islet cell tumors of the pancreas are typically less than three centimeters in size, avidly enhance, and have distinctive hormonally related symptoms.\(^10\) In contrast, the non-syndromic ICTs are usually larger than five centimeters and hypoattenuated in appearance due to their necrotic, cystic nature.\(^10\)

**Diagnosis**

Granulocyte Sarcoma of the Pancreas

**Summary**

In conclusion, it is important to consider granulocyte sarcoma when confronted with a pancreatic mass with associated abdominal adenopathy and splenomegaly in a patient with a known history of AML, especially in the absence of biliary ductal dilatation. Although rare, recognizing this diagnosis as a distinct possibility may assist the pathologist in making the correct diagnosis on biopsy specimens and facilitate appropriate chemotherapy treatment with improvement or resolution of tumor burden.

**References**