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Pancreatic Extra-Gastrointestinal Stromal Tumor: An Unusual Presentation of a Rare Diagnosis

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Background: Gastrointestinal stromal tumors (GISTs) rarely develop outside the digestive tract and such tumors are designated extra-GISTs (EGISTs). The majority of EGISTs are located in the mesentery, omentum, and retroperitoneum, and the primary localization in the pancreas has been reported in only about six cases. We describe a patient with a large metastatic pancreatic EGIST that had metastasized to the liver at time of presentation.

Case: An 84-year-old male presented with worsening confusion and agitation for the past few days. He also reported progressively increasing abdominal distension for the past 3 years, more so in the past few months. He denied any abdominal pain, nausea, or vomiting. He mentioned one episode of melena 2 months ago. There was a history of unintentional weight loss of 30 pounds over the past few months. Review of systems was otherwise negative. Past medical history was significant for diabetes mellitus and lactose intolerance. Pertinent examination findings included a cachectic appearance, altered mentation without any focal neurologic deficit, and marked abdominal distension with dullness on percussion. Investigations were significant for elevated ammonia level (168 ug/dL), AST/ALT/Alk. phosphatase (424/153/102 U/L), and total bilirubin of 1.7 mg/dL. CEA and CA19-9 were within normal limits. Computed tomography (CT) scan of the abdomen showed an extremely large central heterogeneous mass of 34 x 24 x 27 cm replacing the entire pancreatic tissue and multiple hepatic metastases. Subsequently, a CT-guided liver biopsy demonstrated a spindle cell neoplasm with CD117 (c-kit), CD34, and vimentin-positive cells, consistent with liver metastasis from an EGIST. On day 3, he had massive hematemesis, for which he was transferred to the intensive care unit. His condition rapidly deteriorated with hemodynamic instability and further worsening of mental status. After a thorough discussion about treatment options and prognosis, his family concluded to limit care to comfort measures only. He passed away the next day, day 5 of admission. The family refused an autopsy.

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Discussion: EGISTs are usually asymptomatic depending on the location, tumor size, and mucosal involvement. The usual clinical symptoms include abdominal pain, early satiety, ileus, bleeding, anemia, and weight loss. This is the first reported case of pancreatic EGIST presenting with abdominal distension due to massive size of the tumor and hepatic encephalopathy secondary to hepatic metastasis.

GISTs originate from the interstitial cells of Cajal (ICC), which express the c-kit protein (CD117), CD34, and vimentin. An EGIST or metastatic disease may mimic a sarcoma or myoma on gross and microscopic appearance. The definite diagnosis is based on the immunohistochemical examination. CD117 expression is the most sensitive marker, found in 95% of GISTs. Frequent mitotic activity (2/50 HPF), high cellularity, and the presence of necrosis indicate a potentially aggressive clinical course for EGIST. The current definitive treatment for GIST, including EGIST, is surgical. Adjuvant therapy appears to be warranted because of high recurrence rates. Imatinib mesylate, a tyrosine kinase inhibitor, is a current treatment of choice for advanced GIST. It has also proven to be of benefit as adjuvant therapy with surgery to prevent recurrences.