Gastric gastrointestinal stromal tumor and neuroendocrine pancreatic tumor: Always neurofibromatosis?

Dear Editor,

A 52-year-old female patient presented to the emergency department with hematemesis, hypotension and tachycardia. She had a history of hypertension and denied other illnesses. The gastroscopy revealed an ulcerated 35 mm subepithelial lesion in the upper gastric corpus. The computed tomography excluded suspicious lymph nodes and metastasis. On endoscopic ultrasound (EUS) (UCT 10-140 AL5; Olympus) the lesion was hypoechoic and heterogeneous with ill-defined borders [Figure 1a] and originated in the fourth layer. Incidentally, an isoechoic 5.8 mm Doppler negative nodule was identified in the pancreatic tail [Figure 1b]. Fine needle aspiration was taken with a 22C needle from the gastric lesion and with a 25-gauge needle from the pancreatic nodule. Cell block cytology was consistent with a gastric stromal tumor [Figure 2a-c] and a neuroendocrine pancreatic tumor [Figure 3a and b]. The patient was readmitted 6 days after discharge with recurrent bleeding and hemodynamic instability and underwent an atypical gastrectomy. At 2 years follow-up there is no evidence of gastric stromal tumor recurrence nor pancreatic neuroendocrine tumor size increase or metastasis.

Gastrointestinal stromal tumors (GISTs) are rare, representing <1% of all gastrointestinal tumors. Interestingly, 13% of sporadic GISTs occur with a second malignancy, more commonly (47%) gastrointestinal carcinomas, but also neuroendocrine tumors of any location in 3% of cases.[1] In the literature, there are four cases of pancreatic neuroendocrine tumors coexisting with GISTs in patients without type 1 neurofibromatosis (NF-1).[2-5] Interestingly, in NF-1 patients in whom these tumors are frequent, there are only 9 cases reporting this association.[3] The question raised by the present case is whether this association is a coincidental finding or are we actually overlooking the pancreas when assessing GIST patients. A systematic evaluation of the pancreas, when EUS is performed in this setting, could help clarify this question.

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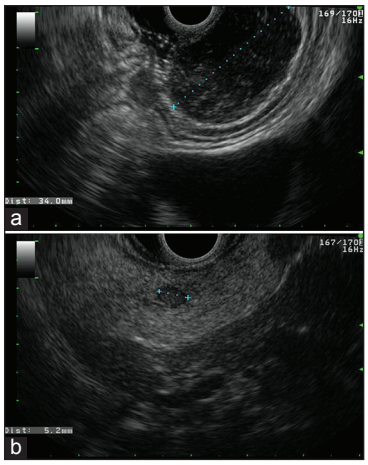

Figure 1. Linear endoscopic ultrasound depicted: (a) 35 mm hypoechoic heterogeneous mass in the upper gastric corpus. Layer or origin muscular propria; (b) incidental 5.8 mm isoechoic nodule in pancreatic tail

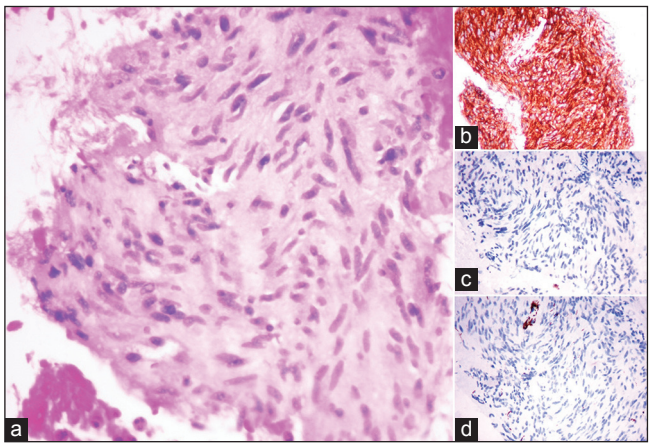

Figure 2. Cytology examination of the gastric mass showing: (a) Spindle cells with elongated nuclei and eosinophilic cytoplasm (H and E stain; ×400); (b) positive staining for CD117; (c) negative stain for S100; (d) negative stain for smooth muscle actin

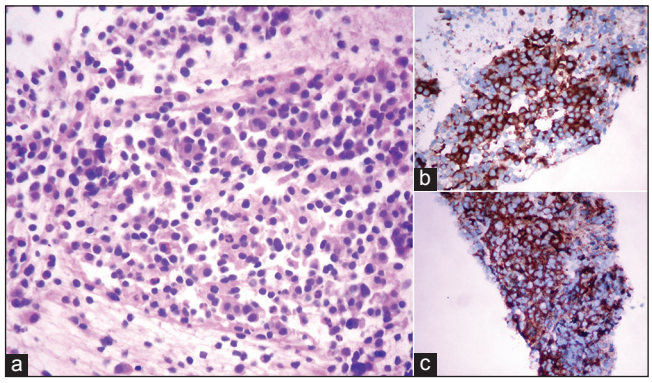

Figure 3. Cytology examination of the pancreatic nodule depicting: (a) Loose aggregates of cells with round and uniform nuclei with fine chromatin and eosinophilic cytoplasm (H and E stain; ×400); (b) positive staining for synaptophysin; (c) positive stain for cromogranine A

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