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CASE REPORT

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Duodenojejunal mass in a patient with neurofibromatosis Type I: A case report

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Case report

AA 68-year-old female with a pertinent past medical history of neurofibromatosis type I and transection of the left ureter was transferred to a regional hospital for recurrent upper GI bleeds and acute on chronic anemia. Computed tomography (CT) enterography was obtained which was remarkable for a large mass in the proximal jejunum and distal portions of the duodenum. Subsequent esophagogastroduodenoscopy (EGD) was performed and random biopsies were obtained which supported a spindle cell tumor. The patient underwent a diagnostic laparoscopy, exploratory laparotomy, proximal jejunal resection of tumor, duodenojejunostomy, and ureteral ureterostomy of the left side due to transection of the left ureter. The patient tolerated the procedure well and her postoperative course was uneventful. Due to gross evaluation and immunoprofile of histological testing gastrointestinal stromal tumor (GIST) was determined to be the etiology (Figure 1 and Figure 2).

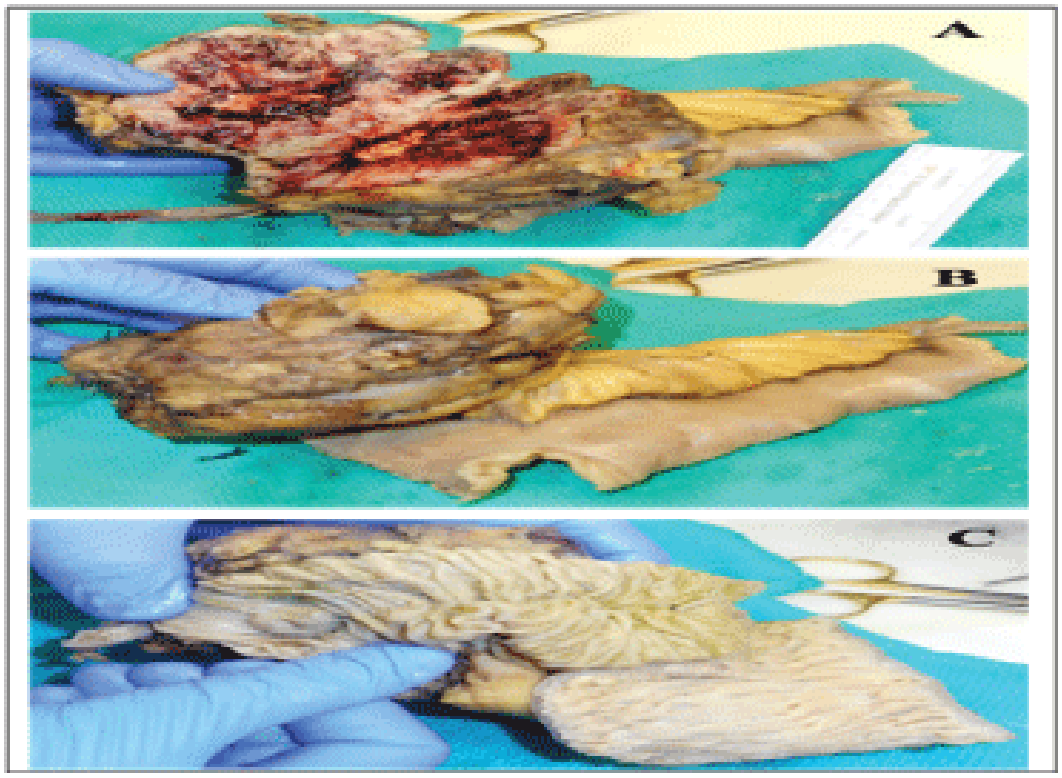

Figure 1. Segmental bowel resection of proximal jejunum with retroperitoneal mass. (A) Grossly, the bowel wall was lined by gray-tan to green-tan, velvety mucosa with a single firm polypoid mass, 1.5 x 1.3 x 0.4-centimeters. (B) The polypoid mass was associated with an intact gray-white firm mesenteric mass, 10.2 x 10 x 6.5 cm. Sectioning revealed a focally hemorrhagic, pale tan, cerebriform cut surface. (C) Non-neoplastic bowel and its associated lumen.

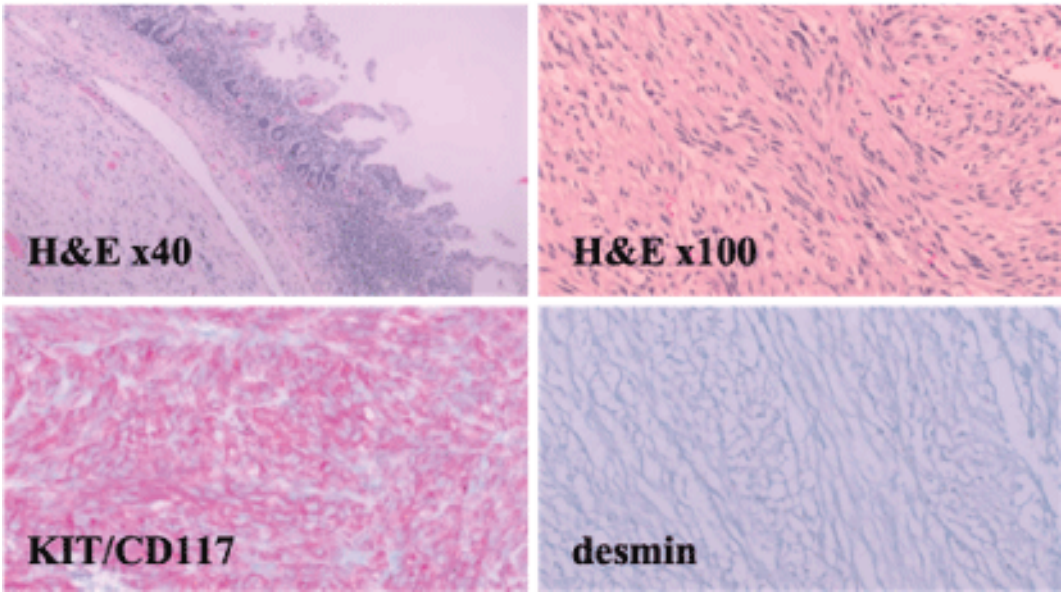

Figure 2. The surgical pathology showed a neoplastic tumor infiltrating the submucosa of eroded small bowel tissue (H&E stain, x40). Higher-power magnification of the neoplastic tissue revealed fusiform-appearing spindle cells with a high cellularity (H&E stain, x100). Properly controlled immunohistochemical staining was performed and showed the tumor cells were strongly positive for KIT/CD117, but were negative for CD34, desmin and S100. Ki-67 staining showed a very low proliferative index. This immunoprofile supported the diagnosis of gastrointestinal stromal tumor.

GISTs are the most common mesenchymal tumor of the gastrointestinal tract. Clinically and oncologically GISTs are diverse. Although they can develop in any segment of

of the gastrointestinal tract, they most commonly involve the stomach [1]. Most commonly GISTs present as small to moderate-sized benign tumors, but uncommonly can metastasize and/or transform into malignant sarcomas [1]. Thus, it would advantageous for radiologists to remain familiar with typical and atypical subtypes of GIST [2]. Histologically, GISTs are organized into three subtypes, with the spindle cell subtype being the most common accounting for approximately 70% of cases [3]. Targeted therapy by tyrosine kinase inhibition to KIT and PDGFRA mutations have revolutionized modern GIST therapy. There are known syndromes associated with GIST tumor development. For example, Carney triad and Carney–Stratakis syndrome, neurofibromatosis type 1, familial GISTs, and desmoid tumor associations [2]. Statistically, when patient's with an underlying diagnose of neurofibromatosis type 1 present with GIST tumors, they are more likely to be of smaller caliber, negative of mutations, and of the histologic spindle type. Uncommonly GISTs metastasize. When this occurs liver and peritoneal involvement are more likely [4]. Moreover, GISTs rarely disseminate outside the abdominal cavity [2]. In conclusion, GISTs are a common benign tumor of the gastrointestinal tract. They are often associated with other syndromes and could possibly present atypically. However, GISTs typically present within the abdominal cavity and remain there. Growth, proliferation, and spread of GISTs can be observed but GISTs rarely metastasize outside the abdomen. As a result, these differences in development, presentation, and progression can have important impacts on evaluation and management.

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