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Case Report

Synchronous Occurrence of Small Intestinal Stromal Tumor and Cecal Adenocarcinoma

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Synchronous development of gastrointestinal stromal tumors with other tumors in the digestive tract is relatively rare, and often occurs in the stomach. We report a 76-year-old woman, who suffered from the exceedingly rare synchronous development of small intestinal stromal tumor and cecal adenocarcinoma. The patient presented with intestinal obstructive symptoms. The pre-operative abdominal ultrasonography demonstrated a very large heterogenous periumbilical mass. Laparotomy was performed and the evaluation of the surgical specimen revealed a huge 14-cm subserosal mass of high risk ileal stromal tumor along with cecal adenocarcinoma with regional lymph node metastasis. The patient died within 3 months of diagnosis.

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Introduction

Gastrointestinal stromal tumors (GISTs) constitute the most frequent group of mesenchymal neoplasms in the gastrointestinal tract.[1] They have been distinctly separated from leiomyomas, leiomyosarcomas, and schwannomas according to clinicopathological differences and immunohistochemical studies.[2,3] They are considered to arise from the interstitial cells of Cajal (ICC) in the digestive tract.[4] Mutations of c-kit proto-oncogene that normally encodes the KIT receptor tyrosine kinase (CD117) in the Cajal cells seem to be responsible for activation of KIT receptor and stimulation of cell growth.[5] Thus, consistent immunohistochemical staining for CD117/KIT is noted in all histologic variants of GISTs.[6]

The most frequent anatomic sites of these tumors are stomach, small intestine, colorectum, and esophagus, respectively.[7] Synchronous occurrence of GISTs with other tumors in the gastrointestinal tract have been reported rarely in the literature. There are some case reports and case series about the coincidence of GISTs with gastrointestinal adenocarcinomas,[1,7] adenocarcinoma arising in villous adenoma of the ampulla of Vater,[8] mucosa-associated lymphoid tissue lymphoma,[9] carcinoid tumor,[7] duodenal Brunner's gland adenoma,[1] neurofibroma, and somatostatinoma of the ampulla of Vater.[10]

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Case Report

A 76-year-old woman was admitted to the emergency room of our hospital, because of progressive periumbilical abdominal pain over the past three days, associated with nausea, vomiting, and obstipation. Her medical, drug, and family history were unremarkable.

On physical examination, the abdomen was distended, having five bowel sounds per minute. Epigastric and periumbilical regions were tender in deep palpation. The vital signs, rectal exam, and other examinations were all normal. Abdominal ultrasonography revealed a very large heterogenous mass in periumbilical and hypogastric regions, surrounded by distended intestinal loops, which were filled by fluid and gas. Celiac or mesenteric lymphadenopathy and ascites were not identified. Liver and pancreas appeared normal. The sonographic impression was a

gastrointestinal neoplasm (Figure 1). Abnormal laboratory findings were as follows: hemoglobin (HB): 10.4 g/dL, mean cell volume (MCV): 76 fl, mean cell hemoglobin (MCH): 25 pg, mean cell hemoglobin concentration (MCHC): 33.8 g/dL, serum glucose: 200 mg/dL, urea: 95 mg/dL, and creatinine: 1.6 mg/dL.

At laparotomy, a huge ileal mass and a tumoral lesion in cecum were identified and partial resection of the ileum along with right hemicolectomy was performed. Macroscopic examination of the ileal segment revealed a huge, grayish-brown subserosal mass, measuring 14×10×8 cm and weighing 700g, located in the mesenteric side of the ileum. The ileal mucosa appeared normal, but the lumen was narrowed due to mass effect. The cut surface of the mass was tan-brown, solid, with hemorrhagic areas.

Macroscopic evaluation of the cecum, demonstrated a white firm, infiltrative tumoral lesion with superficial necrosis, measuring 8×5×3 cm.

Microscopically, the ileal mass was a neural type gastrointestinal stromal tumor, originated from the outer layers of the muscularis mucosa. The tumor was composed of spindle cells showing palisading and whorl-like configurations, with areas of hemorrhage and necrosis (Figure 2). Twelve mitotic figures per 50 HPF (>10/50 HPF) and the large size of the tumor (>10cm), placed this neoplasm in the high-risk category (according to risk categories for GISTs from Fletcher et al.).[11]

The tumor cells were immunoreactive for CD117 (c-kit), vimentin, S-100, and neuron specific enolase (NSE). They were negative for cytokeratin, CD 34, desmin, and actin (Figure 3).

Microscopic evaluation of the cecal lesion, revealed a moderately differentiated adenocarcinoma with extension into the pericolic fat and appendix, vascular invasion, and involvement of six regional lymph nodes. Cecal margin was involved by adenocarcinoma. The patient died within 3 months of diagnosis.

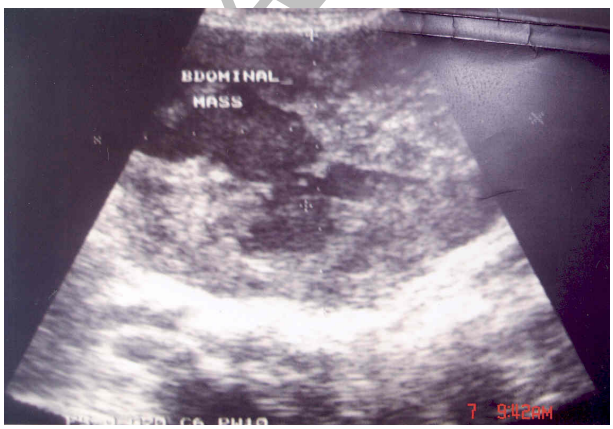

Figure 1. Preoperative abdominal sonography revealed a very large heterogenous periumbilical mass.

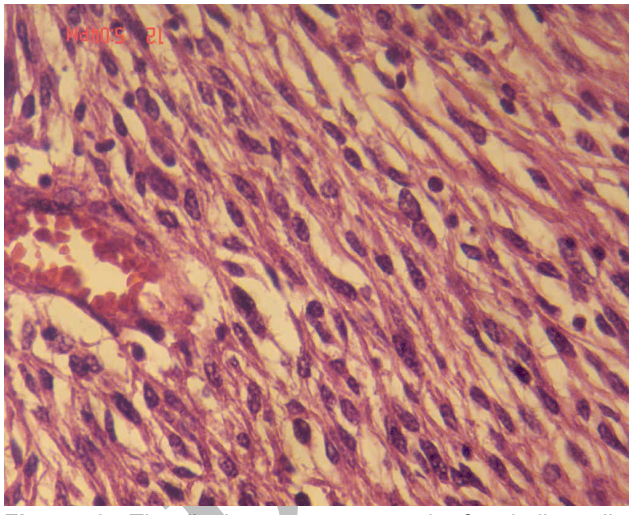

Figure 2. The ileal tumor composed of spindle cell showing palisading and whorl-like configurations.

Discussion

The simultaneous development of GISTs and adenocarcinomas of digestive tract are mainly described as case reports and case series. Most of them have been reported in the stomach.[7,12–16]

In a series of 200 cases of GISTs studied by urbanczyk et al. synchronous tumors were present in seven patients, including one adenocarcinoma of colon.[7]

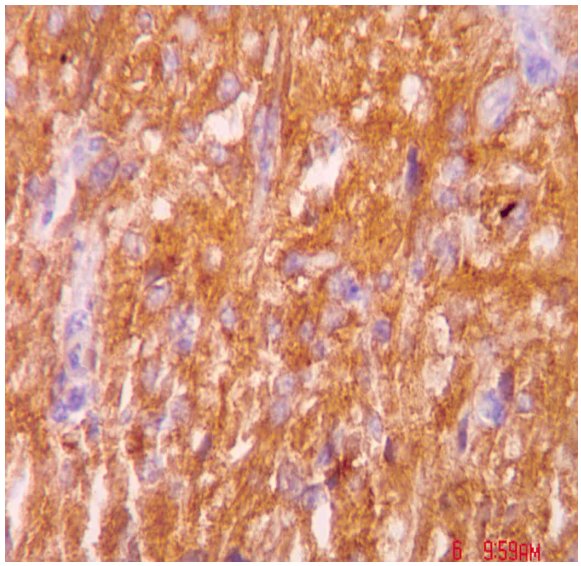

Figure 3. The tumor cells are immunoreactive for CD 117.

In the study by Wronski et al. on 28 patients with GISTs, four gastric stromal tumors (14% of patients) were found with a second neoplasm, including two gastric adenocarcinomas, one colon adenocarcinoma, and a gastric lymphoma.[17] Few reports of synchronous gastric stromal tumor with large bowel and pancreatic adenocarcinoma also exist.[18,19]

Although it seems that the synchronous occurrence of stromal and epithelial neoplasms of gastrointestinal tract is more common than it has been considered, small bowel stromal tumor concomitant with colon adenocarcinoma is virtually rare. A Hungarian study on 43 patients with GISTs, revealed seven patients with a second tumor, including three small intestinal GISTs occurring metachronous or synchronous with colorectal adenocarcinoma.[1] Melis et al. also reported two cases of small bowel stromal tumor with synchronous invasive colon adenocarcinoma.[20]

In the presented case, the cecal adenocarcinoma was clinically silent and the patient's symptoms were mainly due to the mass effect of small intestinal stromal tumor. In a report by Kövér et al., the stromal tumors in four of seven synchronous tumors were silent. This confirms the importance of surgical intra-abdominal control before closure.[1]

Recently, it is believed that the combination of tumor size and mitotic activity is the most important prognostic factor in GISTs.[11] Poor prognosis of this patient is simply explained by the presence of a high-risk GIST and an adenocarcinoma with lymph node metastasis.

The etiology of synchronous gastrointestinal stromal and epithelial tumors is unknown, but some theories exist. In areas with high incidence of gastrointestinal cancers the coincidence alone can easily account for such an association. Otherwise gene mutations might interact with two neighboring tissues inducing the development of different tumors in these areas.[14]

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