CASE SNIPPETS

Dermatofibrosarcoma protuberans of the jejunum

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We report a 55-year-old man with recurrent bleeding from the small intestine. Preoperative investigations suggested it to be a small intestine tumor. The resected specimen was diagnosed at histology as dermatofibrosarcoma protuberans of the small bowel.

[Indian J Gastroenterol, 2001; 20: 30]

Key words: Gastrointestinal bleeding, small intestine tumor

Tumors of the small intestine are rare and sarcomas of the intestine are even rarer. Dermatofibrosarcoma protuberans is a tumor reported to arise from the dermis of the skin. We report a patient with dermatofibrosarcoma protuberans arising from the intestine; such an occurrence has not been described before.

A 55-year-old man presented with six episodes of tarry black stool and three syncopal attacks during these bleeds, in one year. There was no history of fever, intake of NSAIDs or bleeding disorder. Examination revealed mild pallor and no positive finding on abdominal examination.

Upper and lower GI endoscopic examinations were normal. Ultrasonography revealed gallstones and a solid mass of approximately 8-cm size in relation to the intestinal loops. Barium meal follow-through study was normal. CT scan confirmed the ultrasonography findings. Digital subtraction angiography showed increased vascularity in this mass.

Laparotomy revealed a mass of approximately 8 cm diameter on the antimesenteric border of the jejunum about 45 cm from the duodeno-jejunal flexure. There was no mesenteric lymphadenopathy and other viscera were normal. Resection of the bowel with 15-cm margins was done and primary anastomosis was carried out. The patient recovered uneventfully and is on follow up for 5 years without evidence of disease.

Cut section of the tumor revealed a solid mass with a healed ulcer in the center. Microscopic examination showed cellular growth with spindle cells arranged in an interwoven pattern or radiating from a central focal point; at places the cells were lying in parallel rows (Fig). A diagnosis of dermatofibrosarcoma protuberans of the small intestine was made.

Onoda et al. described a 45-year-old man with pigmented dermatofibrosarcoma protuberans (Bednar tumor) arising on the right arm. The tumor recurred twice, with metastases to the skin, lung and brain. The patient succumbed to the disease; autopsy revealed metastases to the skin, lungs, thyroid, pancreas, stomach, small intestine and thigh muscles.

Dermatofibrosarcoma is histologically similar to benign fibrous histiocytoma but grows in a more infiltrative pattern, spreading along connective tissue septa in deep areas. The central portion of the tumor consists of a uniform population of plump fibroblasts arranged in a distinct ordered pattern around an inconspicuous vasculature. Secondary elements such as giant cells, xanthoma cells and inflammatory elements are few or absent altogether. Dermatofibrosarcoma protuberans is a rare tumor arising from connective tissue in the dermis of the skin. In our patient we believe it may have arisen from the connective tissue in the muscle layer of the small intestine. The bleeding was because of ulceration in the central part of the tumor, which is well known in sarcoma of the intestine.

Dermatofibrosarcoma protuberans of the skin is stated to carry a good prognosis. We therefore expect a good prognosis with this tumor arising from the bowel; our patient is well at 5 years without evidence of disease.

References


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Lelomyosarcoma of third part of duodenum with perforation

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Malignant stromal cell tumors of the duodenum present with pain or bleed. We report a patient with lelomyo-
sarcoma of the third part of the duodenum that presented with perforation. [Indian J Gastroenterol 2001;20:30-31]

Key words: Duodenal tumor

Leiomyosarcoma of the duodenum is rare. It occurs in the second part of the duodenum and presents with bleeding or pain. We report a patient with duodenal leiomyosarcoma occurring in the third part of the duodenum, who presented with perforative peritonitis.

A 45-year-old woman presented with history of acute abdominal pain of 3 days duration. There was no significant previous history. The pulse rate was 140/min, respiratory rate 32/min, and systolic blood pressure 90 mmHg. The abdomen was guarded and rebound tenderness was present. Plain radiographs did not reveal free gas under the diaphragm. Ultrasonography revealed free fluid in the peritoneal cavity; CT scan showed retroperitoneal and mesenteric inflammation, in addition to free fluid, with normal pancreas. Seropurulent fluid, aspirated on abdominal paracentesis, showed presence of Gram-negative organisms.

At exploratory laparotomy, the stomach, first two parts of the duodenum, and the rest of the small bowel were normal. There was 500 mL of sero-purulent fluid in the peritoneal cavity. The retroperitoneum was edematous and showed presence of gas; the pancreas was normal. After extensive Kocherization, a large perforation, 1 cm x 2 cm, was seen on the posterior inferior surface of the third part of the duodenum. Palpation revealed a hard mass not involving the serosa, with its distal margin extending into the fourth part of the duodenum. Digital palpation through the perforation revealed complete luminal obstruction by the mass. Multiple mucosal biopsies were taken through the perforation. Resectional surgery was deferred in view of poor general condition of the patient and local sepsis. Diversion was achieved with a T tube, which was inserted through the perforation. A feeding jejunostomy was added. The patient died on the third postoperative day due to septicemia. Histology of the mass revealed epithelioid leiomyosarcoma with spindle-shaped cells and occasional mitosis (Fig).

Leiomyosarcomas of the duodenum are rare, representing only 8%-22% of small bowel malignancies. They commonly occur in the second part of the duodenum.

The growth is usually extrinsic or suberosal. Common presenting symptoms are abdominal pain, weight loss and bleed. We did not find in literature a report of duodenal leiomyosarcoma presenting with perforation. Presentation is usually delayed due to late involvement of the mucosa. Endoscopy and CT scan are useful in diagnosis. Small intestinal barium study and modified duodenography may also be useful. Surgical treatment by duodenocephalopancreatectomy is usually curative for neoplasms in the third part of the duodenum.

The prognosis of duodenal neoplasms is poor because of delayed symptoms, especially in patients presenting with weight loss or obstructing symptoms.

References

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Ganglioneuroma of small intestine presenting with perforation peritonitis

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We report a 42-year-old man with benign solitary small intestinal ganglioneuroma presenting with perforation peritonitis. The patient had no evidence of MEN IIb syndrome. Simple segmental resection was done; the patient is well on follow up one year later. [Indian J Gastroenterol 2001;20:31-32]

Key words: Small intestine tumor

Small bowel tumors constitute only 3% to 6% of gastrointestinal tract tumors. Most of them are found incidentally at operation or at autopsy. Benign tumors may present with pain secondary to intussusception and intermittent incomplete obstruction.

A 42-year-old man presented with diffuse pain in the abdomen and constipation for 2 days, along with fever for five days. The patient was transferred to our hospital from another town. On examination, the abdomen was distended. The patient was pale and jaundiced. Abdominal paracentesis was performed. There was 300 mL of sero-purulent fluid with 7000 white blood cells. The patient had anemia and neutrophilia. A biopsy of the tumor was taken from the distal jejunum. The tumor was 5 cm in size with multiple satellite nodules. The patient was discharged after two days of supportive care.

The tumor was a ganglioneuroma. The patient was asymptomatic on follow up three months later.