Primary Linitis Plastica of the Colon and Rectum

Report of Two Cases

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Primary linitis plastica of the colon and rectum is an uncommon entity. Sixty-six cases have been reported in the English literature. Two new cases are reported, one of the transverse colon with widespread metastases and the other of the right colon extending from the appendix to the distal resection margin of the transverse colon. Some clinical and pathologic characteristics of the tumor are discussed, based on a review of the literature. [Key words: Linitis plastica; Carcinoma; Primary colon; Pathology]

LINITIS PLASTICA is a type of gastric adenocarcinoma characterized by diffuse, rigid thickening of the wall without formation of a localized mass and various histologic and cytologic patterns of carcinoma cells, the most well known of which is signet-ring cell. Primary colorectal adenocarcinomas with the same pathologic features have been sporadically reported.1-18 Ojeda et al. reviewed 52 cases of primary colorectal linitis plastica in the English medical literature and added two cases in 1982.18

We will report two new cases of primary colorectal linitis plastica, one of the transverse colon and the other of the right colon extending from the appendix to the distal surgical margin of the transverse colon.

Report of Two Cases

Patient 1: A 31-year-old Hispanic woman was admitted with complaints of abdominal pain, a 35-pound weight loss, and anorexia for four months, and fever, malaise, nausea, vomiting, and melena for one week. Past medical history was significant for a psychiatric disorder treated with thorazine.

Physical examination revealed a cachectic woman with a temperature of 102°F. The abdomen was moderately distended with a fluid wave. There was left upper quadrant tenderness with a palpable mass and negative bowel sounds. Rectal examination revealed an extrinsic anterior mass and guaiac positive stool. Pelvic examination demonstrated a mass in the cul-de-sac and bilaterally enlarged ovaries. The left lower leg had an ulcer.

Significant blood studies were hemoglobin 10.4 per cent, hematocrit 31.6 per cent, white blood cell count 20.6 cu mm with a left shift, alkaline phosphatase 122.9U, carcinoembryonic antigen 69 ng/ml. The bilirubin, aspartate, and alanine aminotransferase were normal.

Barium enema revealed a stricture in the distal transverse colon that appeared extrinsic. Colonoscopy showed diffusely erythematous ulcerating mucosa to 60 cm and a stricture at that point. Biopsy yielded mucin-producing adenocarcinoma.

Three weeks after admission, the patient was taken for an exploratory laparotomy and an ileosigmoidal bypass was performed, because of a large unresectable tumor mass incorporating the splenic flexure, stomach, and spleen. Ascites and diffuse peritoneal, omental, and organ implants were seen as well. Postoperatively, the patient was given one course of chemotherapy, became neutropenic, septic, and died three weeks later.

Autopsy showed a 22-cm firm gray tan tumor mass encircling the distal transverse colon, rendering it a rigid tubular structure. The tumor extended to and was adherent to the posterior wall of the stomach. It also extended to but did not invade the pancreas (Fig. 1). Cut section revealed tumor replacing the whole wall of the transverse colon up to a thickness of 5.0 cm. The lumen was narrowed to 1.0 cm, and the mucosa was ulcerated and hemorrhagic. There were 3 liters of ascitic fluid, marked adherence of a thickened diaphragm to underlying organs, fibrous adhesions between small-bowel loops, peritoneal and mesenteric implants, and bilateral ovarian enlargement. The liver was normal.

Microscopic examination revealed mucin-producing adenocarcinoma, signet-ring cell type, of the transverse colon with replacement of all three layers of the bowel wall. A few crypts and residual smooth muscle were still evident. Single signet-ring cells were seen, within prominent mucin lakes (Fig. 2), as well as extensive fibrous tissue proliferation. The carcinoma extended transmurally through the posterior wall of the stomach to the submucosa with no mucosal involvement. The tumor also extended retroperitoneally to the pancreas but did not invade it. Metastatic adenocarcinoma was present bilaterally in the ovaries and throughout the peritoneum, including the cul-de-sac.

Patient 2: A 54-year-old white woman complained of increasing difficulty in defecating of two months' duration with no weight loss, anorexia, or melena. She had a long history of vague abdominal right upper quadrant pain.

Physical examination was significant for a benign abdomen and a negative stool guaiac test. The hemoglobin was 13.1 per cent, hematocrit 37.5 per cent. Colonoscopy revealed a stricture at the hepatic flexure, and biopsy revealed "colonic mucosa infiltrated by signet-ring carcinoma."
without ulceration (Fig. 4). Also present were diffuse infiltration of the colonic muscular wall and pericolonic and ileal adipose tissue, with preservation of the wall layers (Fig. 5). There was minimal desmoplastic reaction. All the lymph nodes examined were small but microscopically involved with metastases. The left breast biopsy revealed infiltrating lobular carcinoma with positive estrogen receptors.

Discussion

Liñitis plastica is a type of extensively infiltrating gastric adenocarcinoma. The name was originally used to emphasize its macroscopic similarity to diffuse fibrotic inflammation of a hollow viscus. Unlike conventional carcinoma, which obliterates infiltrated tissue structures, liñitis plastica preserves general demarcation of the three involved layers of the gastrointestinal wall: the mucosa, submucosa, and muscularis propria. Diffuse tubular thickening and rigidity of the wall, the hallmark of liñitis plastica, are results of fibrotic reaction around infiltrating cancer cells i.e., desmoplasia. The same process accounts for induration of adjacent soft tissue infiltrated by carcinoma. Other designations emphasizing this tactile aspect of macroscopic characteristics are scirrhous carcinoma4,7,10 and fibrosing carcinoma.

Gastric carcinoma presenting this form of spread is microscopically a poorly differentiated adenocarcinoma in which acinar arrangement of cancer cells, the histologic hallmark of adenocarcinoma, is abortive at best and the cancer cell population is pleomorphic. A wide range

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**FIG. 1.** (Patient 1). Cross section of the liñitis plastica adenocarcinoma. Note the carcinoma infiltrating the transverse colon (C) with preservation of the muscularis propria, with extension posteriorly to the pancreas (P), and superiorly to the posterior wall of the stomach (S).

**FIG. 2.** (Patient 1). High-power view of the adenocarcinoma. Note the signet-ring cells within a mucin lake (hematoxylin and eosin; × 500).