

PRIMARY LINITIS PLASTICA CARCINOMA OF THE COLON

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THE primary linitis plastica type of carcinoma of the colon is rare. Although David¹ reported an instance of this lesion in 1931, no mention of examination of the stomach was made at the time of surgical exploration. In 1936 Dixon and Stevens² reviewed 37 cases of linitis plastica carcinoma of the colon and presented an additional 6. In all but the case reported by David, the lesion was secondary to gastric neoplasm of the linitis plastica type. More recently, Laufman and Saphir³ have added 4 instances of unequivocal primary carcinoma of the colon of this variety. Necropsy failed to reveal any evidence of gastric carcinoma or other attributable primary sites. All their cases were diagnosed clinically as ulcerative colitis prior to surgery or necropsy; in 2 this disease was an additional finding. A similar lesion has been encountered recently at the Cleveland Clinic Hospital. Clinically and roentgenologically its resemblance to ulcerative colitis resulted in a tenable preoperative diagnosis of this disease. Its true nature was revealed at the time of operation only after the bowel was opened for pathologic examination. Careful exploration of the stomach failed to reveal any abnormality. It appears probable that our case is another instance of primary linitis plastica carcinoma of the colon.

Case Report

A professional baseball player, aged 34, was seen for the first time at the Cleveland Clinic on December 29, 1950, complaining of intermittent diarrhea and symptoms indicating intestinal obstruction. One of us (B. D. R.) had observed this patient at the inception of his disease 20 years previously (1931) when he was admitted to a hospital complaining of bloody diarrhea, malaise, weight loss, abdominal pain and fatigue. The diagnosis at that time was chronic nonspecific ulcerative colitis. Stool cultures were negative for pathogens and no amebae were found. Proctosigmoidoscopic examination was compatible with the above diagnosis. X-rays of the colon were interpreted as follows: "There is no definite evidence of an organic lesion of this colon. The entire bowel is quite irritable and somewhat tender to palpation. The appendix is not visualized."

Except for occasional loose stools the patient was well until 1943 at which time his symptoms returned and he was again hospitalized. Unfortunately, x-rays taken at that

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time are not available for study, but the report from the record was as follows: "A barium enema filled the entire colon and entered the terminal ileum. The colon was spastic in character and showed repeated heavy spasms during filling. There was an absence of haustral markings throughout. A few areas along the descending colon are suggestive of rather large ulcerations. **Impression:** Ulcerative colitis."

In July 1947, the patient again consulted one of us (B. D. R.) because of recurrence of symptoms. He stated that he had been in good health since 1943 and able to play professional baseball until recently. Proctosigmoidoscopy showed the rectal mucosa to be granular and friable. Roentgen films of the colon (fig. 1a) at that time showed early changes suggesting chronic ulcerative colitis, particularly in the sigmoid colon. During 1947 and 1948, he was seen on frequent occasions and treated with sulfasuxidine. Periodic sigmoidoscopic examinations showed typical ulcerative colitis involving the rectum. The patient was not observed again until October 1950 when he experienced a particularly severe exacerbation consisting of eight to ten loose water stools daily, malaise, occasional bright red blood, and weight loss. He stated that deterioration appeared more rapid with this exacerbation than with previous ones.

Physical Examination: The patient was 69 inches in height. Although his usual weight was 240 pounds, at the time of examination his weight was 216. He claimed that he had lost 24 pounds in the previous 6 months. His temperature was 98.6 F., pulse 84 per minute and regular, blood pressure 135/85. His general appearance was satisfactory, although he was somewhat pale. General physical examination was normal. The abdominal musculature was so heavy that satisfactory examination was not possible;

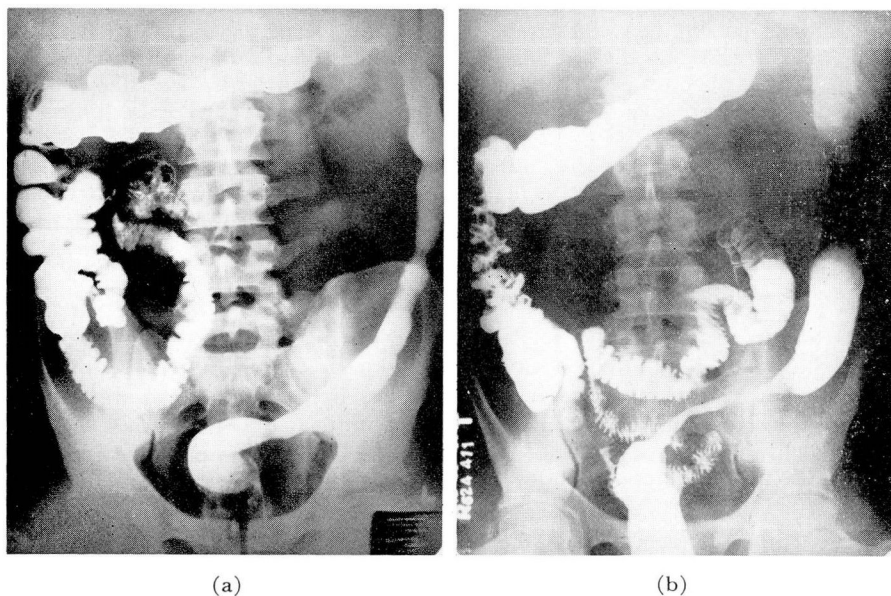


FIG. 1. Ulcerative colitis showing (a) generalized narrowing of the descending and sigmoid portions of the colon with loss of normal haustral markings; (b) colon 3 years and 3 months later with loss of haustrations in transverse and descending portions. Descending portion smooth. Long, narrowed segment involving sigmoid portions represents carcinoma.

however no masses were palpable. A tentative diagnosis of chronic, nonspecific ulcerative colitis was made.

Proctoscopic examination revealed the ampulla of the rectum to be almost obliterated by a stricture at 5 inches; there was evidence of diffuse inflammatory change at this point with profuse bleeding on swabbing. Roentgenograms of the colon (fig. 1b) were reported as follows: "The rectum appears to be of normal caliber. The entire sigmoid portion shows noticeable smooth narrowing for a distance of approximately 16 cm. It appears to be contracted. Descending portion is unusually smooth and there is an absence of normal haustral markings in the transverse portion. Stricture of sigmoid is most likely on the basis of ulcerative colitis rather than neoplasm. Changes in transverse and descending portions are suggestive of ulcerative colitis."

Because the patient continued to decline and because of the rather extreme stricture with obstruction present in the sigmoid colon, surgical intervention was indicated. Accordingly, on January 25, 1951, the left colon was approached through a low mid-line incision, which was extended past the umbilicus to the left costal margin. The sigmoid colon with its mesentery and pericolic fat formed a large mass which nearly filled the pelvis. The descending colon was fixed and leathery and exhibited the hypertrophic changes attendant with chronic low grade obstruction. Transverse colon and cecum appeared normal as was the small bowel. Palpation of the stomach, duodenum and liver revealed no unusual findings. These observations suggested ulcerative colitis with a severe pericolicitis of the sigmoid colon. Because of the localization of the disease, it was decided that the entire left colon should be removed and a splenic flexure colostomy performed in the left upper quadrant. The sigmoid colon was freed from the rectosigmoid upward, the left colon was mobilized in the usual manner, and the splenic flexure was liberated and drawn down to the midline. The colon was divided below through the rectosigmoid and the rectal stump was turned in. The pelvis was re-peritonealized. The proximal colon was divided through the splenic flexure with the Cope-DeMartel clamp, and the abdomen was closed through all layers with vertical steel alloy "figure of eight" sutures, bringing the colostomy out through the midportion of the upper limb of the wound.

The patient had an afebrile, uncomplicated, postoperative course and was discharged from the hospital on the tenth postoperative day. He has since gained 20 pounds and manages his colostomy efficiently enough to permit full time work.

Pathology

Gross Examination: (Fig. 2) The specimen consisted of a segment of descending and sigmoid colon measuring 30 cm. in length. Fifteen centimeters from the proximal line of resection and extending distally for a distance of 14 cm. the serosal surface was dull, gray and indurated as contrasted with the smooth glistening serosal surface of uninvolved bowel. The pericolic fat tissue in the involved region was indurated and greatly thickened. Upon opening the bowel a normal mucosal pattern was apparent in the first 15 cm. from the proximal line of resection and the lumen measured 5 cm. in cross diameter. The mucosal surface of the involved segment was finely granular, firm and gray, without evidence of distinct ulceration. The lumen measured only 1 cm. in cross diameter throughout this region. The wall was thick, gray and firm, converting the involved segment into a rigid tube. On cross section, finger-like projections of dense gray-white tissue extended into the pericolic fat for a dis-



FIG. 2. Gross specimen. Note narrowed segment of bowel due to carcinoma.

tance of 3.5 cm. The mesentery was indurated and greatly thickened throughout. Twenty-nine lymph nodes were recovered from the pericolic adipose tissue and mesentery after alcohol-formalin fixation.

Microscopic Examination: (Fig. 3a and b) Sections of neoplasm revealed absence of normal mucosal glands. A thin layer of fibrinopurulent exudate lined the lumen and extended about the entire circumference of the bowel. Immediately beneath this layer was a malignant epithelial new growth characterized by large and small masses of atypical epithelial cells, for the most part polygonal in shape with round, vesicular nuclei. Atypical mitotic figures and hyperchromatic large nucleoli were numerous. In areas, small lumina were formed by these neoplastic cells. Large and small irregular cystic spaces were lined by these cells and contained blue-staining material. Individual cells lay free in these pools of blue-stained material. An occasional signet-ring cell was observed either alone or in small groups within the tumor stroma which was moderately dense. The signet-ring cell, however, was not an outstanding feature of the lesion. The tumor extended about the circumference of the bowel and invaded all muscle layers, serosa, and pericolic adipose tissue. A mild inflammatory infiltration of polymorphonuclear leukocytes and lymphocytes was noted throughout the lesion. Masses of tumor cells were present in small subserosal venules.

Twenty-eight of the twenty-nine lymph nodes demonstrated secondary carcinoma.

Sections of grossly uninvolved portions of bowel revealed a normal structure without increase in density of submucosa or atypical mucosal pattern.

Diagnosis: Adenocarcinoma of colon; extensive linear and stenosing type — linitis plastica.

Discussion

The clinical and pathologic features of this case simulate those of primary linitis plastica carcinoma of the colon reported by Laufman and Saphir.³ Al-

though ulcerative colitis was an additional finding in 2 of their cases, they were of the opinion that this disease was incidental. The detailed clinical, roentgenologic, and sigmoidoscopic observation of our patient for 20 years indicates

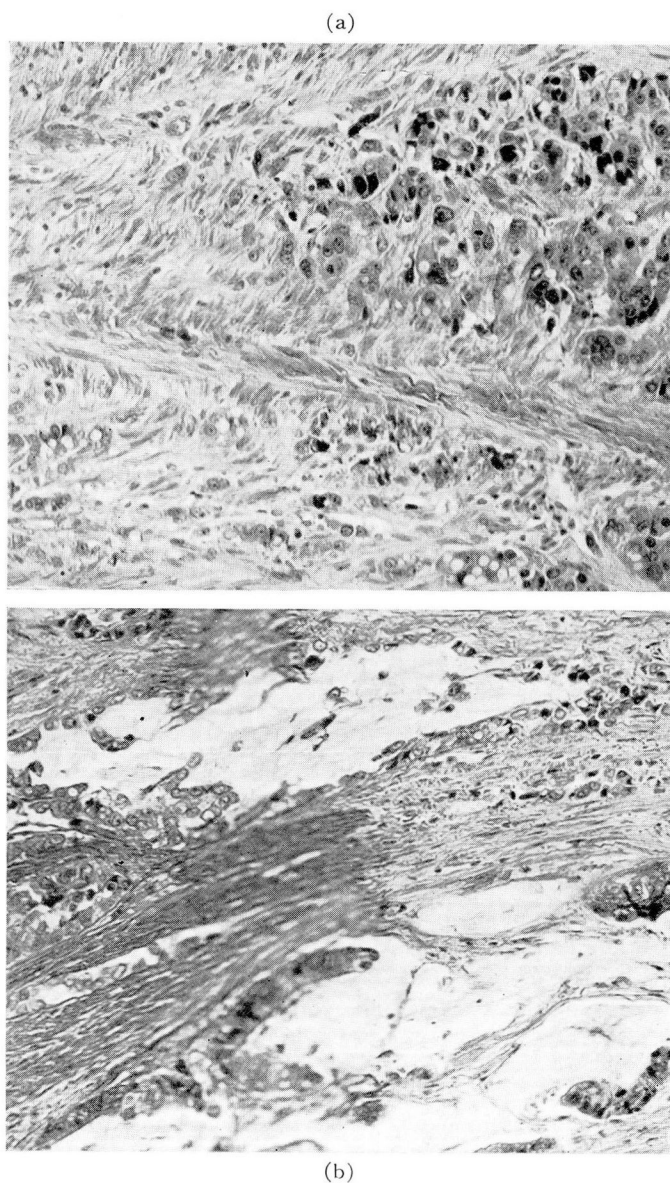


FIG. 3. (a) Photomicrograph showing spheroidal, dark-staining tumor cells in muscularis. Rare signet-ring type present (H & E x70); (b) mucinous pools formed by tumor (H & E x70).

that the initial disease of the intestinal tract was chronic ulcerative colitis. The presence of carcinoma in that segment of previously diseased bowel suggests neoplastic transformation of chronic ulcerative colitis rather than the possibility of coincidence. The involvement of such a long segment of bowel also favors the multicentric origin of this type as stressed by Willis⁴ in his discussion of linitis plastica carcinoma of the stomach.

The diagnostic difficulties encountered in this type of neoplasm present a problem. Clinical, roentgenologic and sigmoidoscopic studies are apparently of little value in the detection of such a lesion arising in pre-existing ulcerative colitis. Patients with similar findings have been followed for years without any indication of neoplastic transformation. Does this imply that they should be subjected to resection of the colon even though their disease appears stationary except for occasional minor exacerbations? Although the rarity of this type of lesion would appear to make this procedure impractical for the group, the fact must be recognized that ulcerative colitis may undergo malignant transformation. Surgical therapy merits particular consideration in those cases in which areas of stricture occur as well as unusual change in a patient's symptomatology.

Primary linitis plastica type of carcinoma of the colon is of additional importance to the surgeon because the presence of such a lesion obviously necessitates more radical surgery than would be undertaken for chronic ulcerative colitis or other benign stenosing lesions of the colon. The validity of the term linitis plastica has provoked criticism and debate. Its entrenched position in medical teaching and literature makes its use imperative in the classification of this lesion. Histologically the tumor herein reported exhibits features attributed to linitis plastica carcinoma as outlined by Laufman and Saphir:³ (1) spheroidal cells, (2) small, miniature new gland formations, and (3) mucin secreting cells. It should be emphasized, however, that many areas of the tumor were similar in histologic appearance to neoplasms frequently encountered in the large intestine not producing diffuse linear involvement.

Willis,⁴ has used the term linitis plastica carcinoma to denote a special macroscopic type of neoplasm. He asserts that histologic variants do not denote entities but merely variants of one entity, while several or all histologic variants may be present in one tumor. Ewing⁵ also stresses the inconsistency of histologic features in those neoplasms which convert the intestine into a rigid tube. It seems more logical to use the term linitis plastica as a gross descriptive one rather than as a connotation for certain microscopic features. The involvement of pericolic adipose tissue, as well as the extensive metastases to lymph nodes, indicates an extremely poor prognosis in this instance.⁶

Summary

1. A case of primary linitis plastica type of carcinoma of the colon arising in a previously existing chronic ulcerative colitis is reported.
2. Preoperative diagnostic difficulties are described.
3. A brief review and discussion of the literature is presented.

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