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Diffuse Lymphoma of Small Intestine and Colon

Case Report with Discussion

S. A. Swenson, Jr, MD* and James L. Omel, MD**

A case is reported of malignant lymphoma of the small and large bowel associated with ulcerative colitis. Primary lymphoma of the gastrointestinal tract is relatively uncommon. The dual association with colitis is rare. Prior to laparotomy, diagnosis of lymphoma is difficult because, of the many possible presenting complaints, none are specific. Treatment is basically surgical excision with postoperative radiation.

This paper adds to current medical literature a case report of malignant lymphoma confined to the gastrointestinal tract in association with ulcerative colitis. The malignant propensity of ulcerative colitis is beyond all doubt. The incidence of carcinoma of the colon in patients with chronic ulcerative colitis is 3% to 5%, or 40 to 50 times that of the general population. This is especially true in cases of long duration. While the vast majority are histologically adenocarcinomas, only a few cases of malignant lymphoma associated with ulcerative colitis have been reported.

Case Report

A 46-year-old white woman had a history of morbid obesity and chronic ulcerative colitis. In April, 1967, when the 5'4" patient weighed 290 pounds, a jejuno-colic intestinal bypass was performed at another hospital.

Four years later, in July, 1971, after losing 156 pounds, the patient complained of blurred vision, dermatitis and edema — signs of vitamin C and A deficiency and electrolyte disturbances. Her intestinal bypass was revised to an end-to-side jejunoileostomy. Several hypertrophied mesenteric lymph nodes noted at operation were submitted for evaluation along with the resected anastomotic site. The pathologic diagnosis was malignant lymphoma, lymphocytic type. The predominant histologic pattern was follicular with extensive involvement of adjacent small bowel submucosa (Figure 1). Liver and spleen were

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noted to be normal at time of the operation. Peripheral blood smears and bone marrow examination were also normal. She was treated with postoperative radiation and progressed satisfactorily except for bowel disturbances alternating between constipation and diarrhea.

In June, 1972, when she was again admitted to the hospital with severe abdominal pain and distension, she was diagnosed as having a volvulus of the sigmoid colon. At operation, there was no evidence of the lymphoma in the liver or spleen. The volvulus was reduced and biopsies were obtained from the colon and small bowel mesenteric nodes. These also contained no residual lymphoma cells. She was placed on prophylactic Cytoxan and Procarbazine chemotherapy.

One year later in May, 1973, she complained of diarrhea and bloody stools. Barium enema revealed very typical generalized ulcerative colitis with megacolon. Treatment consisted of diet and steroid administration.

On September 26, 1973, she was last hospitalized with acute exacerbation of colitis. Admission laboratory studies revealed a hemoglobin level of 11.1 gms with 3.29 red blood cells. Urinalysis was essentially normal. Multiple survey revealed a TSP of 5 gmm%, albumin 3.2%, blood urea nitrogen 7 gmm%, serum bilirubin 0.3 gmm%; alkaline phosphatase 73 international units and serum glutamic oxaloacetic transaminase 23 IU. Serum magnesium was 2.0 gmm% at time of admission. We decided to reanastomose her defunctionalized jejunum and perform a subtotal colecetomy with ileorectostomy. At operation, the edematous megacolon and several huge mesenteric nodes were removed. The liver and spleen were again considered to be normal.

The pathological report indicated severe ulcerative colitis with multiple foci of hemorrhage, hyperemia, and thickened bowel with pseudopolyps. Of even greater significance was the fact that the whole mesentery was a mass of matted lymph nodes which became

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**Figure 1**

Microscopic section of tissue removed July, 1971, at the site of the jejuno-colic anastomosis. (x 260) Lymphocytic tissue is invading the glandular structure of the intestine.
Figure 2
Section from the distal ileum (Sept. 1973) showing lymphocytic tumor invasion. (x 60)
Swenson and Omel

confluent into a huge multiloculated mass adjacent to the ileocecal valve. The tumor extended outside the lymph nodes and involved vessels and adjacent structures. The wall of the distal ileum was also affected (Figure 2). Pathologic diagnosis was malignant lymphoma, lymphocytic type, well differentiated. The sections of large bowel, ileum, and jejunum all had extensive mucosa and submucosal involvement.

A bone marrow examination, performed four days postoperatively, gave essentially normal results. There was no neoplastic involvement and the myeloid erythropoid ratio was a normal 3/1. Some decreased cellularity was noted but this was perhaps due to previous radiation.

The patient recovered from the operation rather slowly. She went home October 24, 1973. She had problems with alimentation and was admitted to another hospital on October 27, 1973. Supportive therapy failed and she expired December 16, 1973.

Incidence

Malignant lymphomas localized to the gastrointestinal tract are not common. In most reports they comprise from 1% to 4% of the malignant neoplasms of the gastrointestinal tract. In a much quoted survey on 618 cases of malignant lymphomas by Gall and Mallory, 11% were localized in the gastrointestinal tract, bone and skin. The stomach is the most common site of primary gastrointestinal involvement, followed by small intestine and colon. A few reports include mention also of esophagus, rectum, and even tongue. Carcinoma is encountered with higher frequency in all sites except perhaps the small intestine. Primary gastrointestinal lymphoma is encountered in all age groups. The peak age for gastric lymphoma is about 50. Small bowel lymphomas predominate in younger age groups. As with all forms of gastrointestinal neoplasms, lymphomas are more common in males than in females.

Ramat has suggested that there is a predominance among underprivileged groups and among some common genetic and environmental factors. He states that intestinal lymphoma is endemic in the Middle East.

Pathology

A lymphoma is a malignant tumor arising from lymphoid tissue, characterized by proliferation and abnormal growth with local invasion and destruction and a propensity to widespread distant metastases and to multifocal involvement. The tumor may arise in almost any organ and may spread to any part of the body. Criteria of primary gastrointestinal involvement used in various studies, as well as the case being presented, include a lymphomatous bowel lesion with only mesenteric lymph node involvement. Other organ structures and peripheral or mediastinal nodes must be uninvolved.

The gross appearance of gastrointestinal lymphoma is often indistinguishable from carcinoma. Tumors may protrude into the bowel lumen forming polyloid masses which tend to ulcerate. They may diffusely infiltrate the bowel wall, causing rigidity or loss of peristalsis. A circumferential infiltration may cause an “napkin-ring” lesion. With extensive involvement of the lamina propria and mucosa, malabsorption can occur — a very common finding. In the early stages of lymphoma the tumor spreads along the long axis of the bowel in the submucosa. As the growth proliferates, ulceration may occur of whatever epithelium is present. By infiltration, the tumor weakens and paralyzes the bowel musculature thus producing dilatation and eventual bowel stasis.

Clinical Features and Diagnosis

The diagnosis of primary lymphoma of the gastrointestinal tract is difficult to establish prior to surgical exploration.

156
Clinically there are many suggestive signs and symptoms but no typical syndrome. Symptoms vary somewhat according to the primary site of involvement.

The most frequently encountered chief complaint is dull abdominal pain. There is usually weight loss with associated malaise, anorexia, and fatigue. Nausea and vomiting are prominent symptoms, the latter often occurring with the onset of colicky pain. A history of altered bowel habits is elicited from the majority of patients. Alternating constipation and diarrhea, as was seen in our patient, is common. A palpable abdominal mass is felt in about one half of the patients. Malabsorption syndrome is very commonly associated with gastrointestinal lymphoma, one series reporting 33% incidence.\(^1\)

Laboratory tests are of little value in establishing the diagnosis. Total and differential white cell counts are only rarely altered. Anemia is also infrequent. Bloody or tarry stools are reported by some patients and up to half have at least occult fecal blood.

The disease may sometimes be suspected on inspection of plain abdominal films since air trapped in the rigid, infiltrated loops of bowel may occasionally delineate the lesions.\(^1\) With colonic involvement barium enema will reveal dilatation of the bowel, segmentation and flocculation of the barium, and thick coarse mucosal folds.

Similar to those reported by others, our patient manifested (a) gastrointestinal symptoms, (b) no superficial lymphadenopathy, (c) no enlargement of mediastinal nodes visible roentgenographically, (d) no hepatosplenomegaly, and (e) normal total and differential white cell counts.

Treatment

In cases where it is technically feasible to extirpate the entire lesion together with its lymphatic drainage, there is general agreement that this should be done. Since many of these lesions at the time of laparotomy may resemble inoperable carcinoma, a frozen section is essential to establish the diagnosis of lymphoma. Postoperative radiation therapy to the tumor bed and areas of secondary spread is the recommended treatment. Chemotherapy is also a valuable adjunct. Survival rates differ according to the site and extent of the initial lesion. In general, small bowel tumors have a poorer prognosis. In a review of 100 cases of primary lymphoma of the gastrointestinal tract, Loehr\(^4\) found the overall five-year survival rate to be 49%. The prognosis is excellent five years after therapy in an asymptomatic patient.

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