Multiple Lymphoid Polyps in Familial Polyposis

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FAMILIAL polyposis and its variant Gardner's Syndrome are well-recognized clinical entities, which generally are readily distinguished from other polypoid conditions of the colon (e.g., diffuse polyposis, juvenile polyposis, Peutz-Jeghers Syndrome, and Turcot's Syndrome) by two striking features.[8] First, they are inherited as autosomal dominant traits with high penetrance and variable expressivity; second, they are considered uniformly fatal by middle-age—generally by age 50—because of adenocarcinoma of the colon. Thus, each offspring of an affected individual has a 50% chance of being affected, and those affected usually manifest the entire syndrome.

This case report describes a patient who had multiple lymphoid polyps of the colon and small bowel in a family with known multiple adenomatous colonic polyposis. Preoperatively, he was assumed to have familial polyposis and thus underwent a total colectomy and ileostomy.

Family History

The family tree of the propositus certainly indicates the presence of familial polyposis (Fig. 1). The patient's mother had colonic polyps and died of colon carcinoma at age 38. The patient's three sisters had total colectomies and ileostomies at the University of Wisconsin Hospitals for multiple adenomastous polyps of the colon. In addition, a maternal aunt and her two daughters are known to have multiple colon polyps. One of these cousins underwent an abdominal colectomy and ileoproctostomy at another institution. No consanguinity is known to exist.

Case Report

A 17-year-old young man was admitted for evaluation as part of a family follow-up for familial polyposis. He had had episodes of abdominal pain for several years. Proctosigmoidoscopic and barium enema x-ray examinations revealed multiple small polyps throughout the colon. Consequently, he was assumed to have familial polyposis and on July 2, 1970, he underwent total colectomy and ileostomy. At laparotomy, the surgeon could not palpate the polyps and therefore performed a colotomy to confirm the presence of multiple small polyps throughout the colon. After the colectomy, the ileostomy was opened and the terminal ileum was carpeted by myriads of 2 to 3 mm. polyps, biopsies of which demonstrated lymphoid polyps. The histologic picture was the same as those polyps within the colon (Figs. 2 and 3). No adenomatous polyps were found in the entire colon. The terminal ileum was not resected to avoid creation of a wet ileostomy.[3, 6, 12] The patient's postoperative course has continued uneventful.

Discussion

Because familial polyposis is considered uniformly fatal, it is generally felt that affected individuals (i.e., those with multiple adenomatous polyps) should be treated in their teens, even when asymptomatic, as malignancy has been reported as early as age 11.[4, 13] Controversy exists as to whether malignancy develops within the colon or

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within the adenomatous polyps themselves. The vagaries and dubious reliability of follow-up and the attendant morbidity of repeated rectal fulgurations have led us to prefer total colectomy with ileostomy over abdominal colectomy with ileoproctostomy in familial polyposis and in Gardner's Syndrome.[9]

Infrequently, adenomatous polyps in this syndrome appear in the small intestine where malignancy is rare. Lymphoid polyps have been reported as multiple polyposis and even as a familial entity.[2, 7] This is the first reported case occurring in a pedigree of familial adenomatous polyposis.

Lymphoid hyperplasia, which includes polyposis, represents a spectrum of disease

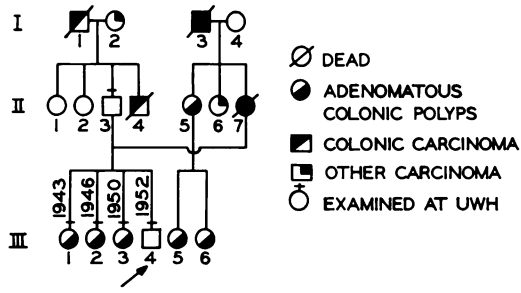

FIG. 1. Family tree with propositus indicated by the arrow.

from diffuse involvement of the entire gastrointestinal tract to isolated polyps.[11] In any of these manifestations lymphoid hyperplasia is considered benign although occasionally surgical treatment is necessary for the complications.[1, 5, 10] Most authors regard

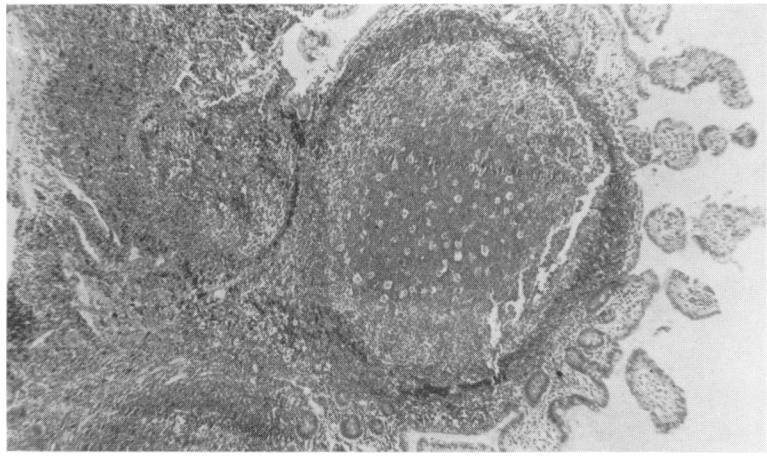

FIG. 2. Focal lymphoid hyperplasia in the mucosa of the terminal ileum to form lymphoid polyps. Magnification ×4.

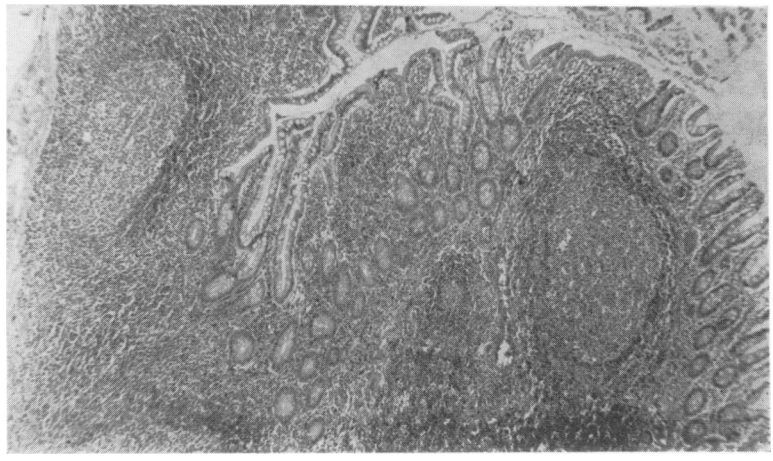

FIG. 3. Similar focal lymphoid hyperplasia in the colon. Magnification ×4.

lymphoid polyps as a benign inflammatory response of lymphoid tissue in the mucosa and submucosa; whereas, they consider adenomatous polyps as a benign epithelial change. The significance of multiple lymphoid colonic polyps in familial polyposis is unknown.

Biopsy is recommended to demonstrate the presence of adenomatous polyps in a patient with colonic polyposis. More cases similar to this are necessary to determine the likelihood of coexisting lymphoid and adenomatous polyps and to establish whether the presence of multiple accessible lymphoid polyps and a strong family history represents another manifestation of familial adenomatous polyposis.

Summary

A case of multiple lymphoid polyposis of the colon and terminal ileum in a patient having a strong family history of multiple adenomatous colonic polyposis is presented. Management of this patient is discussed from the background of these two entities, and a question of their association is raised.

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