Lymphoid Polyposis Associated with Familial Polyposis and Gardner's Syndrome

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This report describes a family with familial polyposis and lymphoid polyps in which a single member exhibits features of Gardner's Syndrome. A review of the literature pertaining to the simultaneous occurrence of lymphoid polyposis with Gardner's Syndrome and familial polyposis is presented. The importance of histologic diagnosis is stressed.

FAMILIAL POLYPOSIS and Gardner's Syndrome are diseases of recognized surgical significance. Both disorders are characterized by a high incidence of carcinoma developing in the large bowel and for which the present acceptable surgical treatment is total colectomy. Lymphoid polyposis, on the other hand, is a benign disorder which may be less familiar to the surgeon. Recognition of the benign nature of this disorder is important in order to differentiate it from familial polyposis and Gardner's Syndrome. Recent reports, however, have noted the simultaneous occurrence of florid lymphoid polyps in patients with familial polyposis and in Gardner's Syndrome. As yet, the significance of this finding is not well understood. This report describes a family with familial polyposis in whom both lymphoid polyps and features of Gardner's Syndrome have been found.

Family History

A family tree of the propositus is presented in Fig. 1. The patients' father had a 20-year history of rectal bleeding and underwent total colectomy for multiple polyposis. An older brother had undergone total colectomy for carcinoma at age 33. Two younger brothers have had total colectomy for multiple colonic polyps. Four other siblings have been studied and no evidence of colonic polyps has been found. A single sibling died in infancy. There is no known history of consanguinity.

Case Report

A 29-year-old Caucasian male was admitted to the Medical University Hospital in 1969 with a history of bloody diarrhea for one year and the recent onset of fever and malaise. His previous history included an operation for peptic ulcer disease four years prior to admission and resection of an abdominal wall tumor the following year. Proctosigmoidoscopy and barium enema revealed extensive colo-rectal polyps which on biopsy were shown to be adenomatous polyps. Subtotal colectomy and ileo-proctostomy were performed in August, 1969. Examination of the surgical specimen revealed a myriad of small adenomatous polyps, a large pedunculated polyp, and a villous adenoma (Figs. 2 and 3). The distal 40 cm of ileum was sigmoidoscoped at surgery and was found to contain numerous small lymphoid polyps. His postoperative course was complicated by intestinal obstruction necessitating re-operation and lysis of adhesions. In 1970, excision of a second abdominal wall tumor was performed and on histologic examination proved to be a desmoid. One year after his colectomy, recurrent symptoms related to the remaining rectal polyps necessitated protectomy and ileostomy which was performed uneventfully. Microscopic examination of the surgical specimen revealed multiple benign lymphoid polyps of the terminal ileum and rectum (Fig. 4). No additional adenomatous polyps were found in the rectal specimen. More recently the patient has had excision of a sebaceous cyst of the right temporal region. Skeletal films have failed to demonstrate any bony abnormalities of the mandible or skull.

Discussion

Multiple lymphoid polyposis of the gastrointestinal tract has been well documented since its original de-

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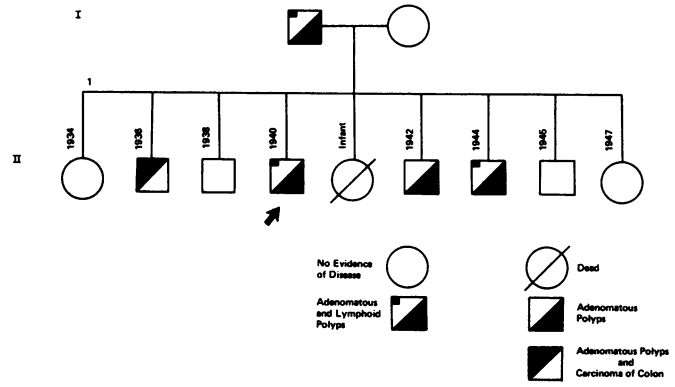

FIG. 1. Family tree with propositus indicated by arrow.

scription by Briquet in 1838.[2,4,14,17] It is now recognized as a benign condition consisting of aggregates of hyperplastic submucosal lymphoid follicles.[2,12] The condition is most frequently recognized in children or adolescents,[1,2] and can be suspected from its radiographic appearance.[15,18] Presenting symptoms are usually mild and consist of alternating diarrhea and constipation with the occasional passage of blood. A tendency toward spontaneous regression of the polyps has been well documented in several cases.[1,2,9] Recognition of this disease by the surgeon is of importance from two standpoints. First, the condition can produce such symptoms as bleeding, intussusception, or intestinal obstruction which necessitate surgical intervention.[13] Secondly, laparotomy may be necessary to differentiate this condition from familial polyposis and Gardner's Syndrome.

The need for histologic confirmation of the diagnosis is especially important since the recognition that benign lymphoid polyposis can co-exist with both familial polyposis and Gardner's Syndrome. In 1968 Thomford and Greenberger reported a case in which lymphoid polyps of the terminal ileum were found at the time of colectomy in a patient with Gardner's Syndrome.[16] A similar finding was reported in two patients with familial polyposis in 1970[3] More recently, another case was reported in which there was a definite history of familial polyposis, but at the time of colectomy, only lymphoid polyps were found.[6]

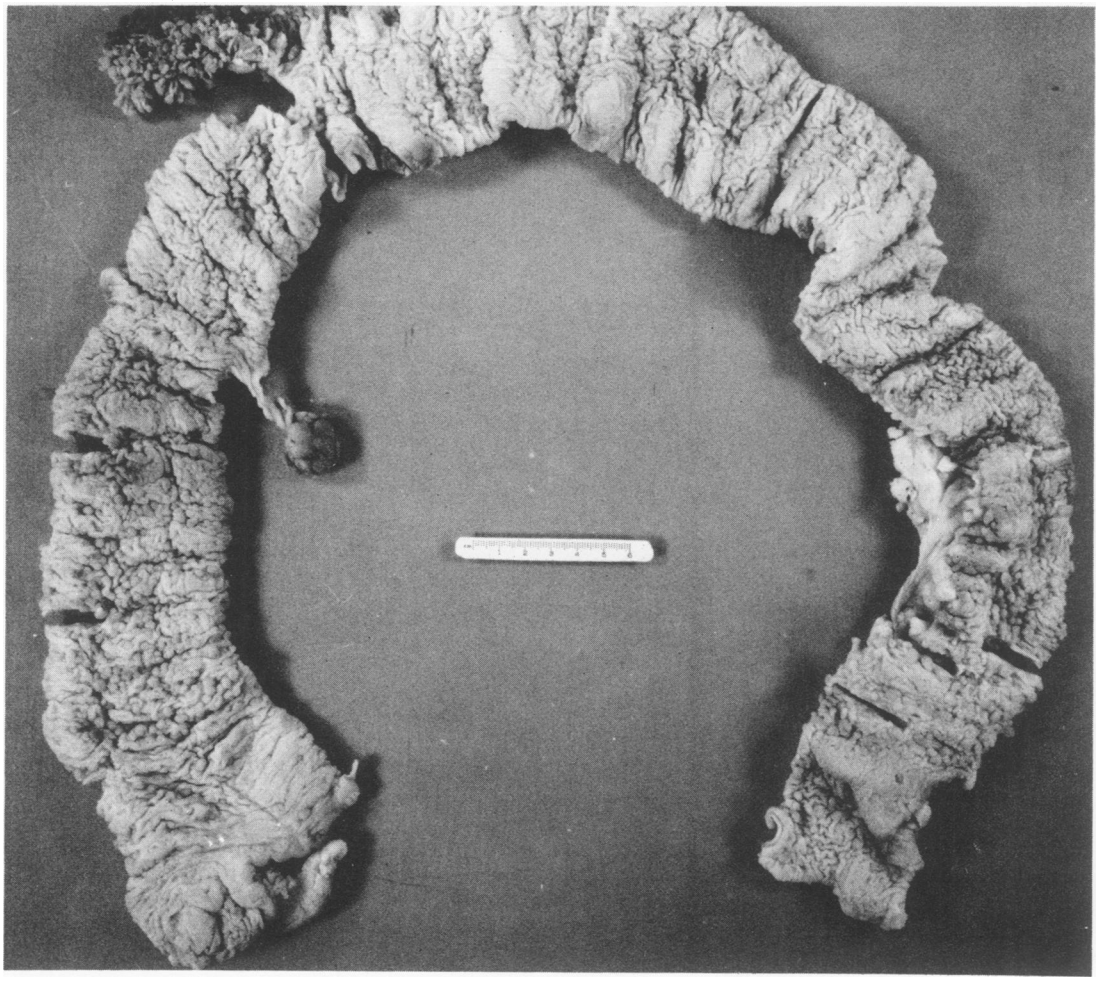

FIG. 2. Subtotal colectomy specimen showing small adenomatous polyps, a large pedunculated adenomatous polyp and villous adenoma. Several small lymphoid polyps can also be seen in the terminal ileum.

Several of the family members in our pedigree had lymphoid polyps of the terminal ileum in addition to adenomatous colonic polyps (Fig. 1). However, the finding of lymphoid polyps and features of Gardner's Syndrome in the propositus is extremely interesting. Familial occurrence of lymphoid polyposis has been described,[2,9] but the simultaneous occurrence of familial polyposis and Gardner's Syndrome in the same family has not been previously reported.[10] Both are inherited in an autosomal dominant fashion and are thought to be controlled by separate genes, although the possibility of the genes being allelic has been considered.[11]

The significance of simultaneous occurrence of lymphoid polyposis of the bowel with both Gardner's Syndrome and familial polyposis is not well understood at present. Gruenberg and Mackman suggest that multiple lymphoid polyps may represent another manifestation of familial adenomatous polyposis.[6] Isolated case reports of carcinoma and lymphoma occurring in association with lymphoid polyposis and dysgammaglobulinemia have been reported.[7,8] One author speculates that specific immunologic abnormalities may be responsible for infection by oncogenic viruses.[7] At present, additional case reports and research are necessary before any possible relationships can be established.

Conclusion

Lymphoid polyposis of the gastrointestinal tract is of importance to the surgeon because it can produce symptoms which require surgical intervention. It may also present as a polypoid disorder requiring differentiation from familial polyposis and Gardner's Syndrome. Rarely, lymphoid polyps may occur simultaneously with familial polyposis or Gardner's Syndrome. Histologic confirmation of the diagnosis of lymphoid polyposis is essential before the appropriate therapy can be provided.

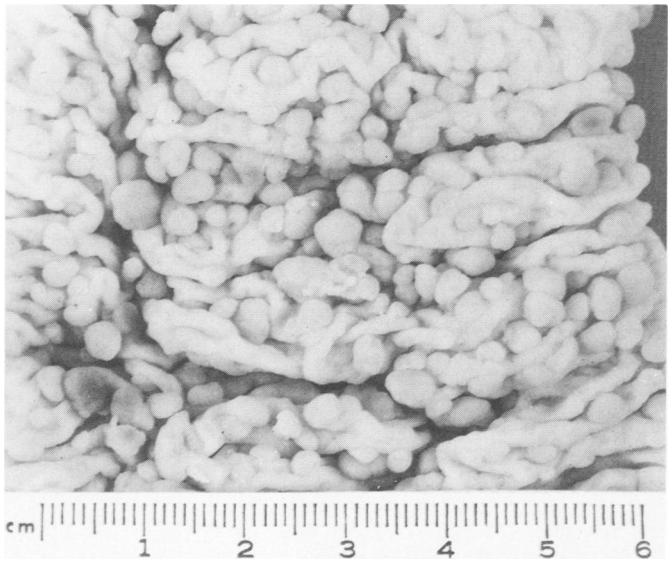

FIG. 3. Closeup photograph illustrating myriads of small adenomatous colonic polyps.

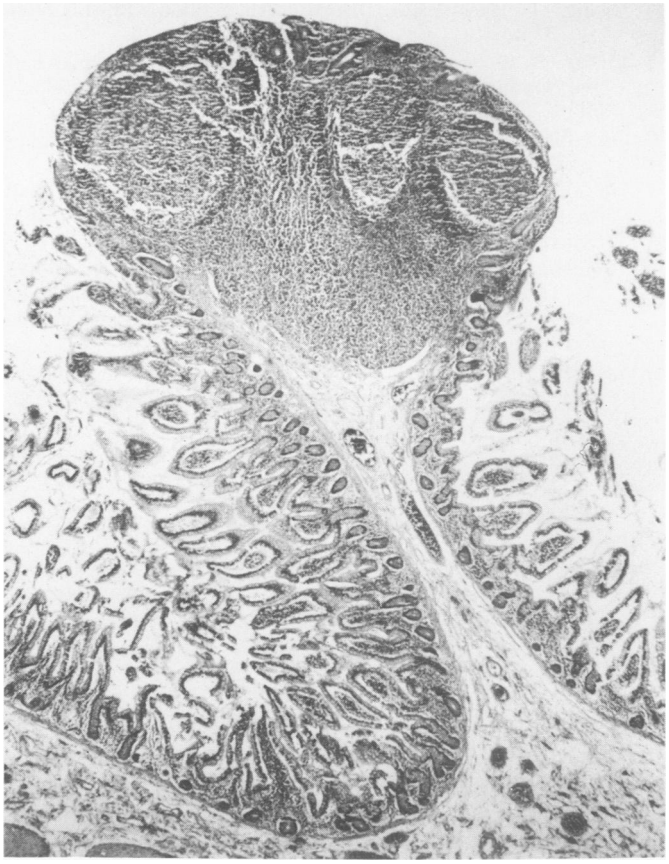

FIG. 4. Microscopic photograph of lymphoid polyp in the colon (×10).

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