Colorectal Lymphoid Polyposis in a Child

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Abstract = Lymphoid polyposis is a lymphoid hyperplasia of the gastrointestinal tract that usually presents as multiple small polyps in the colon during childhood. This should be differentiated from other neoplastic or familial polyposis of the intestine. We report a case of benign lymphoid polyposis of the colon in a 17-month-old boy who presented with perianal fistula and mucosal ulceration. Colon study and rectal examinations showed multiple polyps in the sigmoid colon and rectum.

Segmental resection of the sigmoid colon and rectum showed over 100 small (3-7 mm) sessile or pedunculated polyps that were diffusely scattered throughout the removed segment. The polyps consisted of mature lymphoid tissue with numerous germinal centers, that was located mostly in the lamina propria and submucosa.

Key words: Lymphoid polyp, Polyposis, Childhood, Intestine

INTRODUCTION

Lymphoid polyposis is a part of the spectrum of lymphoid hyperplasia of the gastrointestinal tract and can occur in a family affected by familial polyposis of the colon (Venkitachalam et al., 1978). Although several cases of histologically benign lymphoid intestinal polyposis has been reported to accompany lymphomas or leukemias (Ikeda, 1931) lymphoid polyposis remains as a benign entity. Therefore it is important to recognize this entity for the differential diagnosis of polypoid lesions of the bowel. The lesion occurs in childhood and it is a self-limited disease which should be managed conservatively.

We present a case of colorectal lymphoid polyposis in an infant complicated with perianal fistulas.

CASE REPORT

A 17-month-old boy was admitted to Seoul National University Children’s Hospital because of persistent perianal fistulas and mucosal ulceration in the rectum. He was born normally without perinatal problem. The birth weight was 4.0 kg. The perianal fistulas were first noted at age of 2 months when his mother noted loose stools on several occasions. He was admitted to a local clinic, where he underwent colostomy with the impression of Hirschsprung’s disease.
Fig. 1. The radiographic examination of the colon after barium enema reveals multiple filling defects (arrows) indicative of polyposis.

Fig. 2. Notice numerous small sessile polyps without active inflammatory change or ulceration.

with perianal fistulas. He was transferred to Seoul National University Children’s Hospital at the age of 8 months, where the colon study and rectal examination revealed numerous small polyps in the colon and rectum, suggestive of familial polyposis (Fig. 1). The lumen of the colon was not distended. No family history of colonic polyposis was elicited. The rectal examination of the elder brother of this patient failed to disclose any polyp. The rectosigmoidoscopic examination of this patient revealed multiple small polyps scattered diffusely in colon and rectum. Endoscopic biopsy revealed chronic non-specific inflammatory cell infiltration in lamina propria and goblet cell depletion. All routine laboratory data were within normal limits. He underwent a segmental resection of the colon and rectum. At operation, numerous small polyps were found scattered in the sigmoid colon and rectum.

Pathologic Findings: Fresh specimen consisted of segments of sigmoid colon and rectum, measuring 7 cm in length and 2.5 cm in diameter, and 4 cm in length and 2 cm in diameter, respectively. The internal circumference averaged 6 cm. The mucosa presented innumerable round to oval, sessile or pedunculated polyps ranging from 3 to 7 mm across (Fig. 2). The polyps were scattered through the sigmoid colon and rectum, measuring about 100 in number. There were neither active inflammatory change nor gross ulceration except for one area of hemorrhage. The colonic wall was not thickened and the serosa was smooth and glistening.

Microscopically, multiple sections revealed that the polyps consisted in large part of hyperplastic lymphoid follicles with prominent germinal centers in mucosa and submucosa (Fig. 3). The lymphoid follicles were mostly in the propria and were associated with minimal inflammatory cell infiltration. Immunohistochemical study of lymphoid infiltrates using monoclonal antibodies to T and B cells showed B cells in germinal centers and some T-cells in mantle area. The polyps were covered with normal mucosa and lacked of the cellular atypism and proliferative activity. Many of the polyps developed a short pedicle. Chronic inflammatory cells were seen only in lamina propria but not in submucosa and muscle layer. The muscle layers were not hypertrophic and had normally distributed and Meissner’s plexus. There was no significant increase of vascularity in any of the layers of the colonic wall.
DISCUSSION

Benign aggregates of hyperplastic lymphoid tissue are occasionally encountered in various tissue or organs including the gastrointestinal tract. Mongenot et al. (1985) described incidence of one case of lymphoid polyp out of 183 colonoscopic biopsies carried out in children. Briquet has been credited for the first description of this condition in 1839. Thereafter, Cohnheim presented another case of lymphoid polyp, and proposed the term gastrointestinal pseudoleukemia (Cohnheim, 1865). In 1966 Fieber and Shafer could collect eight cases of pathologically verified lymphoid hyperplasia of the terminal ileum (Fieber and Shafer, 1966). Collins et al. described benign lymphoid polyposis as a form of benign lymphoid hyperplasia. They reported gradually enlarging aggregates of lymphoid tissue in the submucosa, many of which were pulled out on short pedicles by peristaltic action (Collins et al., 1966).

Unfortunately, in the literature the term "lymphoid polyposis" has been used by various authors to describe a malignant condition, and this created some confusion. It could easily be confused with lymphomatous polyposis of the intestinal tract, which includes lymphosarcoma, Hodgkin's disease, reticulum-cell sarcoma and giant follicular lymphoma (Cornes, 1961). Thompson referred the lymphomatous polyps in older patients to be fewer in number and larger (0.5-5 cm), and malignant. In this context, lymphoid polyposis should be distinguished from lymphomatous polyposis because the condition to which we refer occurred in an infant who had diffusely scattered small polyps (3-4 mm) that were histologically benign and, on follow up, had a benign course.

The exact etiology of this condition is not well understood. Early investigators considered it to be a benign variety of lymphoma (Symmers, 1946). Others suggested a chronic inflammatory origin. Some authors thought it represented an intermediate stage between acute mesenteric lymphadenitis or regional enteritis and giant follicular lymphoma (Collins et al., 1966). Venkitachalam et al. described a multiple lymphoid polyposis occurring in a family affected by familial polyposis of the colon and suggested a genetic factor in this condition (Venkitachalam et al., 1978).

Present case would support the possibility of reactive hyperplasia based on histologic and immunohistochemical examinations that showed features of reactive hyperplasia rather than neoplastic proliferation of lymphoid tissue.

Clinically, the patients having lymphoid polyposis have intermittent abdominal pain, diarr-

Fig. 3. The polyps show aggregations of lymphoid follicles in mucosa and submucosa
rhea, constipation, rectal bleeding and intestinal obstruction (Swartley et al., 1962). These cases have been mistaken for acute appendicitis, intussusception, Meckel’s diverticulitis and Hirschsprung’s disease (Fieber and Shaef er, 1966). The cases associated with hypo- and dysgammaglobulinemia and resultant recurrent infection, and giardiasis have also been mentioned (Hermans et al., 1966; Hodgson et al., 1967; Peterson, 1957). This patient was presented only with perianal fistulas. We could find no case in the literature that is complicated with perianal fistula in this condition.

The lymphoid polyposis in childhood is self limited. In most uncomplicated cases conservative treatment should suffice. This child is doing well several years after the surgery up to this time of report.

Our case indicates that lymphoid polyposis should be listed as a differential diagnosis of colonic polyposis in childhood. And for the definitive diagnosis polypectomy should be done. In our case reoperative colonoscopic biopsy showed features of chronic bowel disease. When we reviewed the specimen there was no lymphoid follicles. However, we believe that it is important to recognize the entity of benign lymphoid polyposis before we suggest the diagnosis by biopsy.

REFERENCES

Cohnheim JF: Ein Fall von Pseudoleukemie. Virchows Arch Pathol Anat 1865;33:451-4
Cornes JS: Multiple lymphomatous polyposis of the gastrointestinal tract. Cancer 1961;14:249-57