Case Report

JUVENILE POLYPOSIS COLI WITH ATYPICAL FEATURES

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ABSTRACT

A 22-year-old woman was found to have 25 polyps involving the rectum and the entire colon. Pathologic examination revealed numerous polyps that were predominantly of juvenile type with hyperplastic, adenomatous, villous and tubulovillous features in addition to a villous adenoma. These observations emphasize the relationship between these polyps and the neoplastic potential of juvenile polyposis. Additionally, this is the first case report of juvenile polyposis in association with cervical rib.

Key Words: Juvenile polyposis, Adenomatous polyps, Hyperplastic polyp.

INTRODUCTION

Juvenile polyposis coli (JPC) was first described by Coleman and Eckert in 1956 (1) and later by McColl et al in 1964 (2). It refers to a rare condition characterized by numerous juvenile polyps in the large intestine and less frequently in the stomach and/or small intestine (3,4). Familial and nonfamilial forms of JPC have been described, both of which can be associated with congenital defects, the latter being more frequent (2,5,6-8,9-12).

A remarkable feature of JPC is the wide range of morphological heterogeneity including focal hyperplasia, focal to extensive adenomatous changes that can be found in individual juvenile polyps. In addition, the presence of tubulovillous and villous adenomas of the colon have been described in some cases of JPC (3,7,13-21). Furthermore, colorectal cancer was reported in this form of polyposis (6-8,14,16-18, 22) as well as other intestinal cancers not only in the patient but also in family members (11).

Here we report a variety of morphological atypical features present in most of the polyps removed colonoscopically in a case of JPC, thus presenting futher evidence for and emphasizing the precancerous potential of the disorder. Furthermore, this is the first report of cervical rib in association with JPC.

CASE REPORT

A 22-year-old woman with a history of hematochezia since childhood was referred to for colonoscopy. A single contrast barium enema was reported to reveal multiple filling defects throughout the colon consistent with polyposis. Upper gastrointestinal radiological examination including enteroclysis for the small bowel was normal.

On physical examination a bony mass was detected in the left supraclavicular region which was diagnosed as a cervical rib radiologically. There was no evidence of growth retardation or nutritional deficiency. Blood hemoglobin and hematocrit values were within the normal range. Colonoscopic examination up to cecum revealed 25 polyps ranging 0.3-3 cm in diameter scattered throughout the colon. Most of the polyps were pedunculated some of which were lobulated. Twelve polyps were removed endoscopically in four sessions during the one year follow up. Sessile small polyps were removed with hot biopsy forceps. Family history was negative for gastrointestinal polyposis or cancer. Relatives were not available for examination.

PATHOLOGICAL EXAMINATION

Twelve polyps were removed and examined both macroscopically and microscopically. They ranged from 0.3 to 3 cm in diameter. The larger polyps were grossly lobulated whereas the smaller ones had smooth, shiny surfaces. On section, the cut surfaces of a few polyps were characterized by several mucus containing cysts. On microscopic examination only two polyps of smaller size (less than 1 cm in diameter) were typical juvenile polyps with cystically dilated glands lined by mucus secreting epithelium and abundant inflamed stroma. Their surfaces were partially eroded (Fig.1)

The remaining 10 polyps displayed a variety of atypical features: the larger polyps (more than 1 cm in diameter) had a different morphology whilst retaining some of the features of the juvenile polyp. The surface epithelium was intact, multilobular in outline and covered by a single layer of columnar cells, mostly mucus secreting (Fig.2). The glands were moderately packed and arranged in an irregular pattern. They differed greatly in size and shape, some of them were cystically dilated. The epithelia were characterized by increased stratification, crowding of cells, reduction in mucin, slightly pleomorfic and hyperchromatic nuclei with the presence of mitotic figures suggestive of mild dysplasia. The stroma was moderate in amount and contained scattered lymphocytes, plasma cells, eosinophil and neutrophil leukocytes. Another feature of these adenomatous areas was the conspicious presence of Paneth cells (Fig.3). Ten of

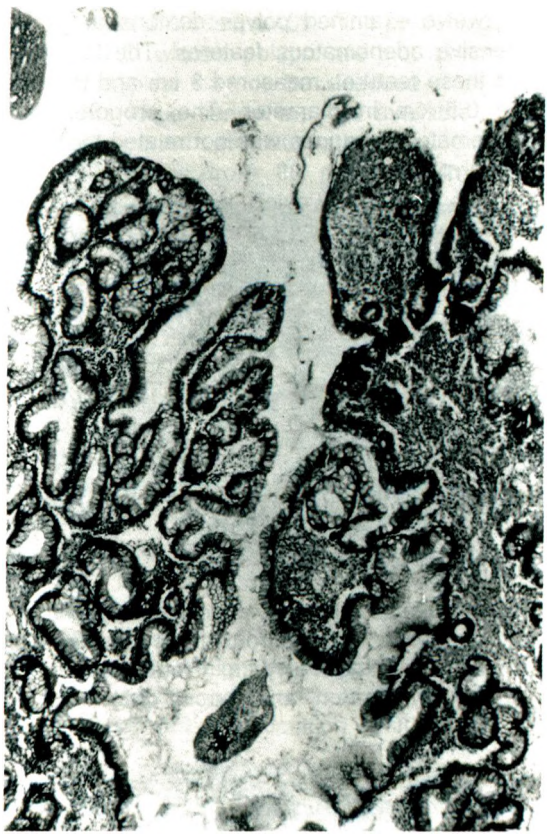

Fig.2.: A polyp with an intact surface epithelium and lobulated contour, closely packed and irregular glands. (H.E. x 40)

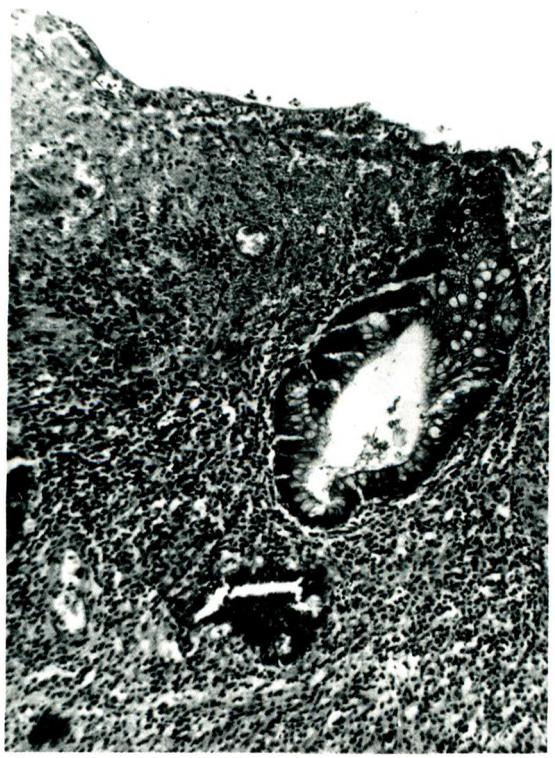

Fig.1.: A portion of a juvenile polyp removed from the splenic flexure has an ulcerated surface and a dilated gland in an abundant inflamed stroma. (H.E x 100)

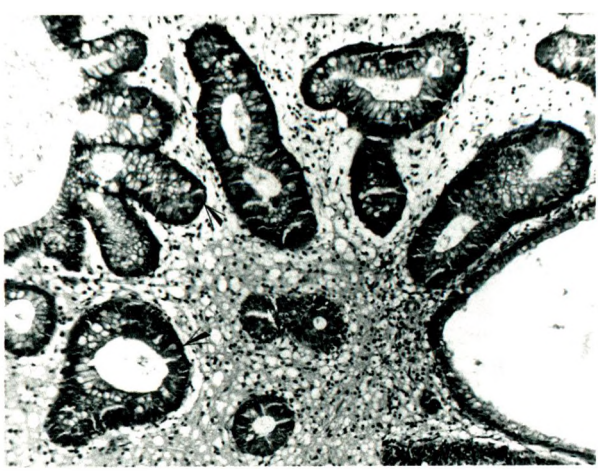

Fig.3.: The presence of Paneth cells (arrows) in juvenile polyp with adenomatous features. (H.E x 100)

the twelve examined polyps demonstrated focal or extensive adenomatous features. The largest polyp with these features measured 3 cm and the smallest one 0.5 cm in diameter. The proportion of the adenomatous changes was not related to the size of the polyp.

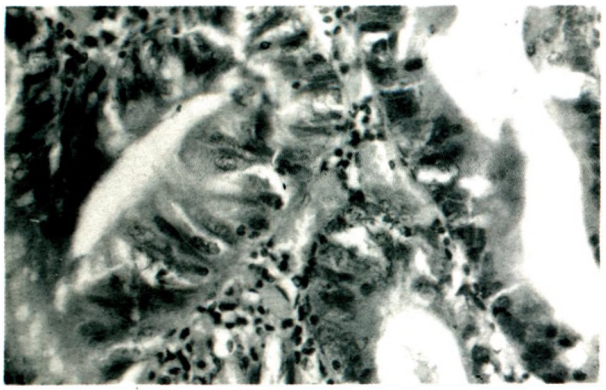

Fig.4.: Glands showing moderate dysplasia in a polyp with tubulovillous features. Cells exhibiting atypical features, increased stratification and mucin depletion. (H.E. x 400)

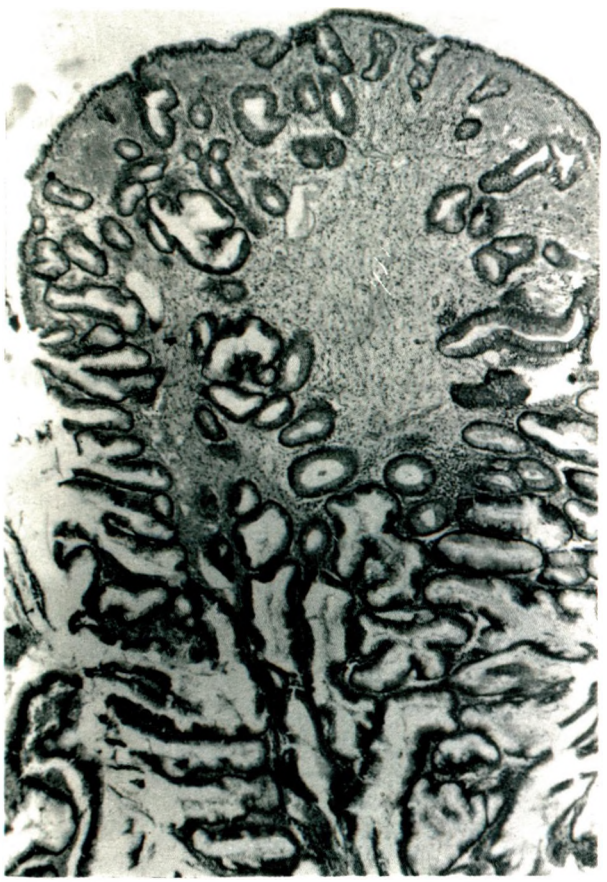

Fig.5.: The tubulovillous adenoma from the splenic flexure. (H.E. x 40)

A pedunculated polyp, removed from the hepatic flexure and 3x2x1 cm in size, showed evidence of moderate dysplasia (Fig.4) in addition to tubulovillous features (Fig.5). Multiple biopsies taken from sessile polyps revealed various histological lesions, including tortuous or racemose glands lined by Goblet cells and

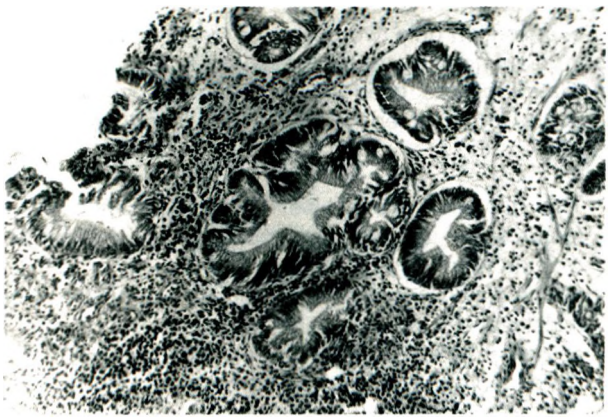

Fig.6.: A portion of a polyp removed from the rectum revealing racemose pattern in a group of glands. (H.E. x 100)

Fig.7.: The villous adenoma from the transverse colon with fingerlike projections lined by mucin secreting epithelium. (H.E x 40)

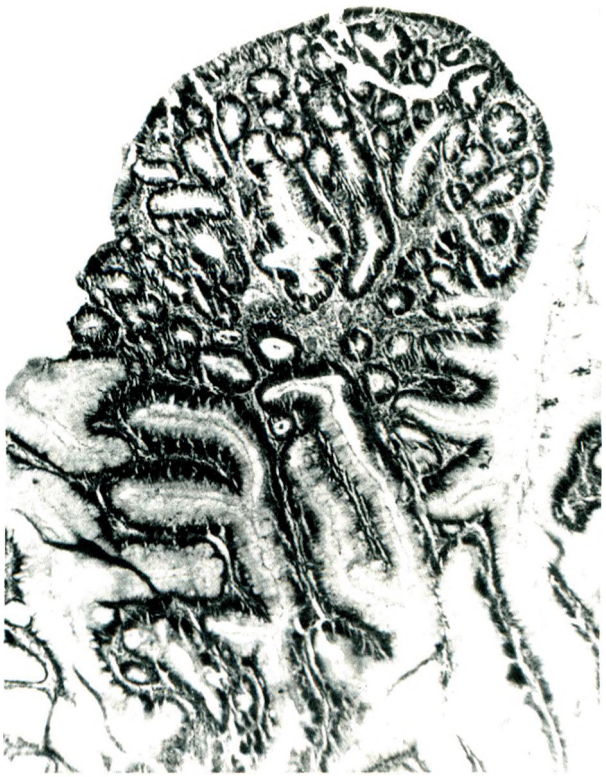

Fig.8.: A portion of the villous adenoma revealing some features of a juvenile polyp. Several dilated glands seen in a wide inflamed stroma. (H.E x 40)

tall, clear columnar cells (Fig.6). In some areas intraluminal papillary projections were noted.

Another pedunculated polyp removed from the transverse colon had a diameter of 3 cm and showed slender villi with tall mucus secreting epithelium lying directly above fingerlike projecting stroma and was diagnosed as a villous adenoma (Fig.7). This adenoma contained focal areas of slightly dilated glands with wide stroma and retained certain features of a juvenile polyp (Fig.8).

No evidence of severe atypia and/or submucosal invasion was found in any of the removed polyps.

DISCUSSION

This case is an example of the morphologically heterogenous nature of the polyps found in JPC. Most of the removed polyps (83%) were atypical in nature. Since care was taken to remove polyps larger than 0.5 cm in diameter, it is probable that the remaining polyps were not altogether atypical in nature. Nevertheless, even if it is assumed that all of the remaining polyps are typically juvenile in character, the proportion of atypical polyps in this case is 40%. The percentage of cases with JPC whose polyps are not uniformly juvenile but atypical cannot be deduced from the literature since large series are not available. The largest series reported upto date is the retrospectively studied 1032 polyps from 80 patients with JPC registered in the St. Marks' Polyp Registry Study (21). Twenty percent of these polyps did not confirm to the classical description of juvenile polyps. Information about the proportion of atypical polyps in a given patient with this disorder is not available.

Epithelial dysplasia is reported in 50% of the atypical polyps whereas only 9% of typical juvenile polyps show dysplasia (19). In our case only one polyp with tubulovillous features revealed moderate degree of dysplasia.

Some of the patients with JPC have congenital defects (2,5,9,11-13). To the best of our knowledge the presence of cervical rib has not been reported in JPC previously. Since cervical rib is a common congenital defect, the association might be coincidental; although it is possible that this congenital abnormality might be genetically determined in association with JPC.

The polyps in JPC or rarely isolated juvenile polyps may show various morphological features besides their usual histology. Hyperplastic (15-17,19,20), adenomatous changes (4,11,16) and villous foci (7,19) have been described as well as villous adenomas (8,22), tubulovillous changes (7,11,16) and adenocarcinomas (6,7,16,17,19). It appears that cases of JPC with various morphological patterns have increased malignancy potential in contrast to those without these features (6,7,16.17,23,24), therefore deserve careful follow up.

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