Virchows Arch. A Path. Anat. and Histol. 382, 355–360 (1979)

Virchows Archiv A

Case Report

Colorectal Polyposis With Mixed Juvenile and Adenomatous Patterns

Guido Monga[1], Gianna Mazzucco[1], Francesco Paolo Rossini[2] and Franco Presti[2]

[1] Istituto di Anatomia e Istologia Patologica dell'Università di Torino (III Chair), Torino, Italy
[2] Divisione di Gastroenterologia, Ospedale San Giovanni Battista, Torino, Italy

Summary. An unusual form of colorectal polyposis is described displaying juvenile, adenomatous and mixed patterns in a 17-year-old girl. Although juvenile polyposis is generally considered to be non-neoplastic in nature, in both the present and in other case reports histological findings support a neoplastic nature. Since an increase in the incidence of large bowel carcinomas has been found in subjects with a previous diagnosis of juvenile polyposis, these patients should be considered to be at risk, and submitted to follow up.

Key words: Colorectal polyposis – Juvenile polyps.

Introduction

Juvenile polyposis is considered to be a different entity from familial polyposis (McColl et al., 1964; Veale et al., 1966). Several cases have been described in the last decade (for a general review, see Enterline, 1976). Morson and Dawson (1972) suggested that there may be two different types of juvenile polyposis, one non-familial and associated with other congenital abnormalities, such as cardiac defects or hydrocephalus; the other characterized by autosomal dominant inheritance (Erbe, 1976), without associated congenital abnormalities. In juvenile polyposis, single lesions display the typical histological features of juvenile polyps (Morson and Dawson, 1972); they have been described in the large bowel only (Veale et al., 1966; Smilow et al., 1966; Stemper et al., 1975) or in the large bowel and in other parts of the gastrointestinal tract (Sachatello et al., 1970; 1974). These polyps are generally considered to be hamartomatous and therefore non-neoplastic (Morson and Dawson, 1972), although this has been questioned since adenomatous areas have been occasionally found (Kaschula, 1971; Koruka, 1976; Enterline, 1976). We report here a case of juvenile

Send offprint requests to: Dr. Guido Monga, Istituto di Anatomia e Istologia Patologica, Via Santena 7, I-10126 Torino, Italy

0340-1227/79/0382/0355/$01.20

polyposis presenting with adenomas, and polyps showing intermediate (juvenile and adenomatous) features.

Case Report

A 17-year-old girl, complaining of intermittent diarrhea and rectal bleeding from the age of six, was referred to the hospital because of anemia, weight loss and severe diarrhea with blood and mucus in the stools. Colonoscopy showed multiple polyps, 0.5 to 2.5 cm in diameter: they were numerous in the rectum and less so in the caecum and ascending colon. A few others were scattered in the left colon and sigmoid. The patient did not show evidence of the Peutz-Jeghers syndrome. X-ray of the stomach and small intestine were normal. Polypoid lesions of the rectum, sigmoid and left colon were removed endoscopically and examined histologically. Ten months later, a right colectomy was performed and 16 polyps were studied histologically. On further sigmoidoscopic examinations from 4 to 10 months after colectomy, 4 more polyps were removed. Fourteen months after colectomy the patient is doing well: she has recovered from bloody diarrhea, anemia and gained weight. Her family history was reviewed and 13 members of the kindred were considered. None of them complained of gastrointestinal pathology; three of them were dead because of acute vascular accidents.

Results

Several sections of 86 polypoid lesions were examined histologically. Particular attention was paid to the amount of stroma, the degree and type of inflammation, the characteristics of superficial and glandular epithelium.

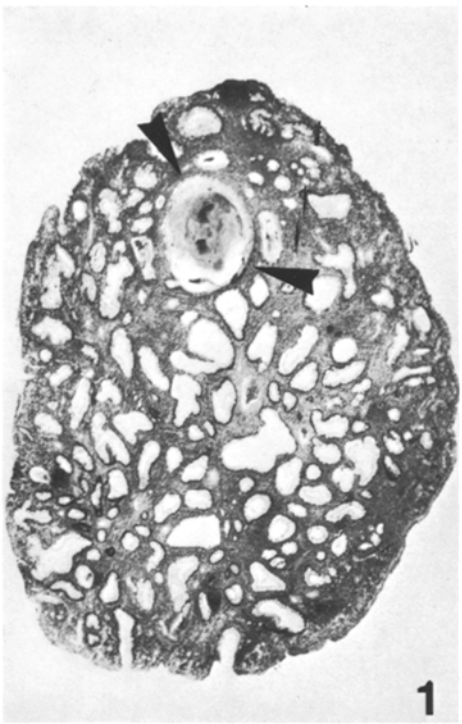
More than half the polyps displayed the typical features of juvenile polyps (Fig. 1): the lamina propria was abundant and oedematous with prominent inflammatory infiltration of both granulocytes and lymphoid cells. Extensive areas of haemorrage were evident. Glands were widely separated by lamina propria and some had a cystic appearance. The cysts were filled with mucus and their epithelium was often atrophic or missing. The surface of the polyp and the glands were covered by a single layer of columnar cells showing no evidence of crowding or anaplasia; most of them were secreting mucus.

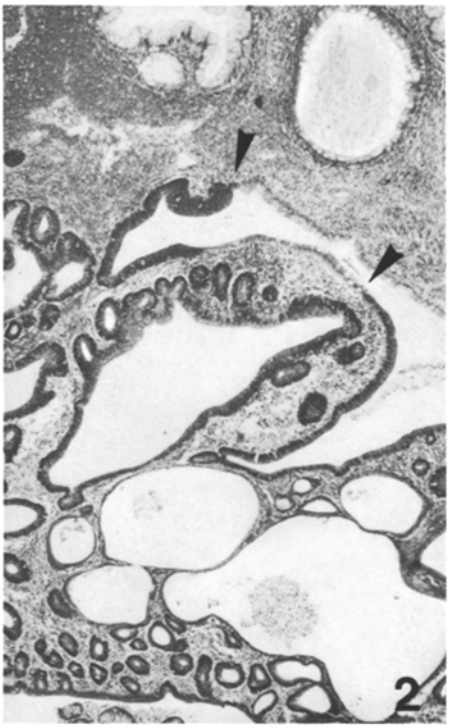
In 15 other polyps features suggestive of adenoma were found in some areas (Fig. 2). Glands were more closely packed, with an irregular outline; epithelial cells were tall, with crowded and irregular nuclei. The number of goblet cells was reduced.

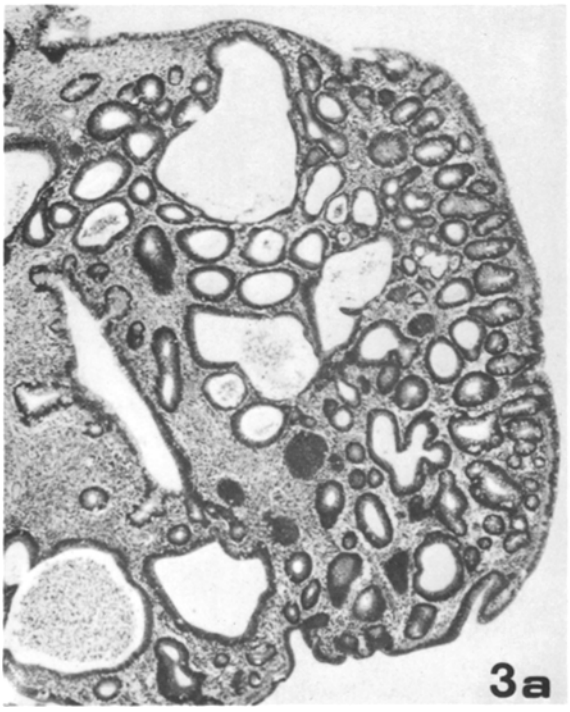
Fig. 1. Juvenile polyp. The surface is regular, the superficial and glandular epithelium typical. Note a retention cyst *(arrows)*. The stroma is abundant and dissociate by haemorrage. ×10

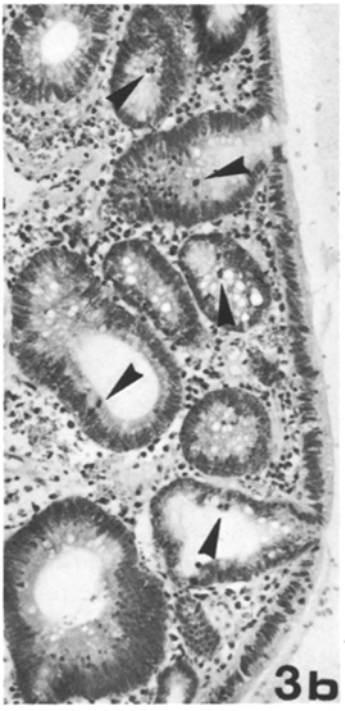
Fig. 2. Polyp with intermediate (juvenile and adenomatous) features. On the top typical glands and a retention cyst are evident. In the middle and on the bottom the glandular epithelium show deeply stained, crowded nuclei. Arrows point out the transition from normal to neoplastic epithelium. ×35

Fig. 3. a Polyp with adenomatous pattern of the glands. Note the abundance of the stroma, similar to that of the juvenile polyp. ×35. **b** Higher magnification of a tubular adenoma. Glandular epithelium show deeply stained, crowded nuclei. Numerous mitoses are evident as well *(arrows)*. ×150









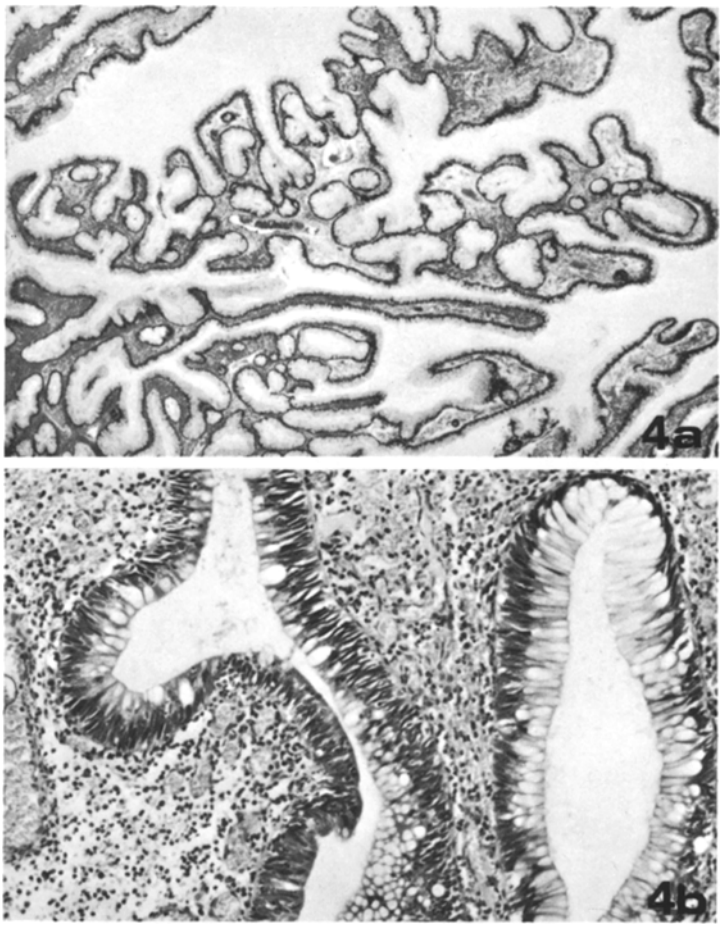


Fig. 4. **a** Part of the villous adenoma with prominent haemorragic dissociation of the stroma. ×10. **b** Higher magnification of the villous adenoma. In an oedematous and congested stroma two glands are present, one (*on the right*) covered by typical mucus-secreting epithelium the other by a neoplastic epithelium with mild displasia. ×250

Finally, epithelial arrangement typical of adenomas was found in 14 additional polyps, being of tubular type in 13 (Figs. 3a and b) and of villous type in one (Figs. 4a and b). Epithelial cells showed prominent proliferative activity, loss of polarity and mild to moderate degree of dysplasia (Fig. 3b). Nuclei were crowded and occupied most of the total cell height. Mucus secretion was inconspicous, goblet cells being rare or absent. Supporting stroma was more abundant than usual in the adenomas and showed inflammatory cell infiltration and prominent haemorragic disruption (Fig. 3a). In the villous adenoma cellular atypia was mild an mucin secretion was abundant. Stromal haemorrage was evident (Figs. 4a and b).

There was no evidence of atypia suggesting malignancy or of submucosal invasion in any of the lesions.

Discussion

The present case has some peculiar features uncommon in juvenile polyposis. The family history rules out an inheritance of the Mendelian type. In addition, in contrast with most cases of the non-familial type of polyposis there were no associated congenital abnormalities. Although the polypoid lesions of juvenile polyposis are usually considered to be non-neoplastic (Morson and Dawson, 1972) our case presented with tubular, villous and other adenomas with intermediate features, apart from the juvenile polyps.

Intermediate juvenile and adenomatous patterns in the same polyp (Kozuka, 1976) and the concomitant presence of a juvenile polyp and an adenoma (Stemper et al., 1975) have been reported. Since both lesions are common their association may be fortuitous. Two cases similar to ours have been reported in the literature (Kaschula, 1971; Enterline, 1976).

These findings suggest a possible relationship between juvenile polyps and adenomas. In this context patients with juvenile polyposis have been described in kindreds with adenomatosis of the large bowel (Veale et al., 1966; Haggitt and Pitcock, 1970). It has been suggested that juvenile polyps are an immature variant of adult (adenomatous) polyps and that they are able to develop into carcinoma, after a considerable period of time and repeated exposure to carcinogenic agents (Kozuka, 1976). An increased incidence of colorectal carcinoma in patients with juvenile polyps or polyposis has been reported (Smilow et al., 1966; Stemper et al. 1975). It is known that juvenile polyps even though they occur in adults are much more common in children and adolescents. Adenomas do not usually appear before the age of 10 and adenomatosis coli is usually detected in the third and fourth decades (Morson and Dawson, 1972). It is therefore conceivable that the neoplastic transformation may occur in the period of life covered by the reports of Kaschula, and Enterline and ourselves, i.e., between 11 and 17 years.

In conclusion, our findings seem to give further support to the possible association of juvenile and adenomatous polyposis and to the possibility of a transformation of the former into true adenomatous neoplasia. Since the development of carcinoma from adenomas is well established, these patients must be considered to be at risk. A careful follow-up may be recommended.

Acknowledgement. The authors are indebted to Miss Clara Bassi, Mrs. Maddalena Gastaldi and Mr. Mario Mariani for the technical assistance.

References

Enterline, H.T.: Polyps and cancer of the large bowel. In: Current topics in pathology, vol. 63: Pathology of the gastro-intestinal tract, B.C. Morson (Ed.), p. 95. Berlin, Heidelberg, New York: Springer 1976

Erbe, R.W.: Inherited gastrointestinal-polyposis syndromes. New Engl. J. Med. **294**, 1101–1104 (1976)

Haggitt, R.C., Pitcock, J.A.: Familial polyposis of the colon. Cancer **26**, 1232–1238 (1970)

Kaschula, R.O.C.: Mixed juvenile, adenomatous and intermediate polyposis coli. Report of a case. Dis. Colon Rectum **14**, 368–374 (1971)

Kozuka, S.: Nature of juvenile polyps in the large intestine. Acta Pat. Jap. **26**, 500–518 (1976)
McColl, I., Bussey, H.J.R., Veale, A.M.O., Morson, B.C.: Juvenile polyposis coli. Proc. Roy. Soc. Med. **57**, 896–897 (1964)
Morson, B.C., Dawson, I.M.P.: Gastrointestinal pathology. Oxford-London, Edinburgh, Melbourne: Blackwell Scientific Publications 1972
Sachatello, C.R., Pickren, J.W., Grace, J.T. Jr.: Generalized juvenile gastrointestinal polyposis. A hereditary syndrome. Gastroenterology **58**, 699–708 (1970)
Sachatello, C.R., Hahn, I.S., Carrington, C.B.: Juvenile gastrointestinal polyposis in a female infant: report of a case and review of the literature of a recently recognized syndrome. Surgery **75**, 107–114 (1974)
Smilow, P.C., Pyror, C.A.Jr., Swinton, N.W.: Juvenile polyposis coli. Dis. Colon Rectum **9**, 248–254 (1966)
Stemper, T.J., Kent, T.H., Summers, R.W.: Juvenile polyposis and gastrointestinal carcinoma. A study of a kindred. Ann. Int. Med. **83**, 639–646 (1975)
Veale, A.M.O., Mc Coll, I., Bussey, H.J.R., Morson, B.C.: Juvenile polyposis coli. J. Med. Genet. **3**, 5–16 (1966)

Received January 2, 1979