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Juvenile Colorectal Polyposis in Children: About a Clinical Case

S.Roudi[1*], Z. Benjelloun[1], A. Ait Errami[1], S. Oubaha[2], Z. Samlani[1], K. Krati[1]

[1]Departement of Hepato-Gastro-Enterology, Arrazi Hospital, Mohammed VI university hospital center, Marrakech 40000, Morocco
[2]Departement of Physiology, Faculty of Medicine, Cadi Ayyad University, Marrakech 40000, Morocco

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***Corresponding author:** S.Roudi
Departement of Hepato-Gastro-Enterology, Arrazi Hospital, Mohammed VI university hospital center, Marrakech 40000, Morocco

Abstract **Case Report**

Juvenile polyposis syndrome is a rare autosomal dominant hereditary disease characterized by the presence of several juvenile polyps in the gastrointestinal tract. This disease predisposes to colorectal cancer, hence the importance of an early detection and a rigorous endoscopic screening [1]. We report the case of a juvenile colorectal polyposis in a 16 years old child revealed by a chronic rectal bleeding.

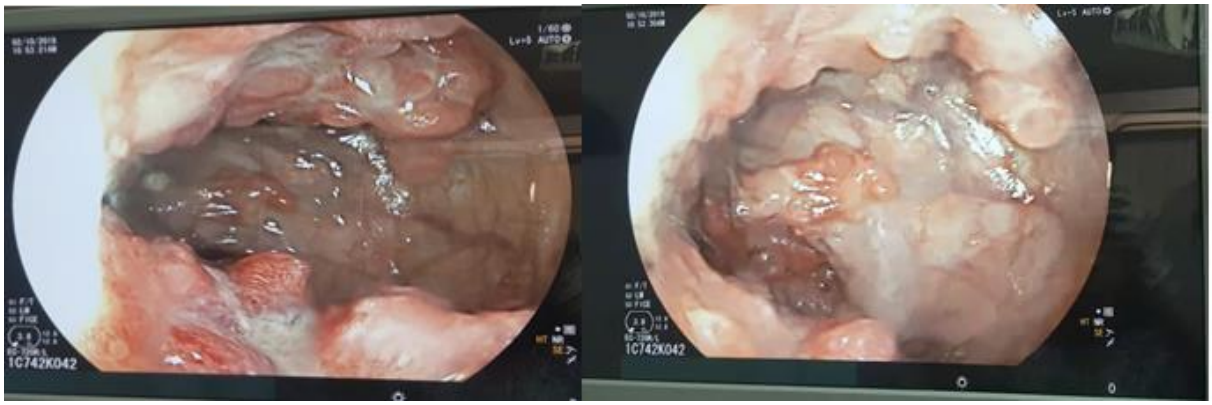
Keywords: Juvenile polyposis, hamartomas, colorectal cancer, endoscopic screening.

INTRODUCTION

Juvenile polyposis is a rare hereditary disease characterized by the presence of multiple hamartomatous polyps in the gastrointestinal tract known to be at risk of malignant degeneration. The most suggestive symptom is rectal bleeding and the histological study of the polyps is crucial for the positive diagnosis of the disease [2]. The therapeutic attitude depends on the number of polyps and their histological characteristics. We report the clinical case of a 16-year-old child whose diagnosis of juvenile colorectal polyposis was revealed during a workup for chronic rectal bleeding.

CASE REPORT

This is a 16-year-old child with a personal history of operated coarctation of the aorta and no known history of familial polyposis. The patient consulted for intermittent chronic rectal bleeding evolving since 5 years. The clinical examination revealed a 2.5 cm long non-stenosing process, bleeding on contact, 3 cm from the anal margin. A total colonoscopy was performed and revealed a colon full of sessile and pedunculated polyps of variable size with the presence of a large polypoid mass 3 cm from the anal margin, non stenosing, friable and bleeding after contact. The histological study of the colonic polyps was showing a juvenile polyp with low-grade and high-grade dysplasia in the rectal polypoid process estimated at 5%.



Figures 1 and 2: Rectal polypoid process, non stenosing with the presence of numerous other sessile and pedunculated polyps with raspberry –like surface

Because of the large number of polyps in the colorectal area, the histological character which shows dysplasia, a surgical treatment was decided. The patient had a total coloproctectomy with ileo-anal anastomosis.

Esophagogastroduodenoscopy did not reveal any other polypoid location. After 2 years, the patient is doing well and the family investigation did not find any other case of juvenile polyposis.

DISCUSSION

Juvenile polyposis is a rare hamartomatous polyposis: the incidence is estimated at 1/100,000 births in Europe and the USA [3]. It is an autosomal dominant inherited disease whose genes are located on chromosome 18 (SMAD4 gene) and chromosome 10 (BMPR1A gene) and more recently mutations of the BMPR1A/ALK3 gene have been identified [5]. The disease affects both genders and usually begins between the age of one and seven years olde but can occur at any age [6].

Diagnostic criteria for juvenile polyposis include: the presence of more than 3-5 colonic juvenile polyps or by juvenile polyps disseminated throughout all the gastrointestinal tract or by at least one juvenile polyp in a patient with a family history of juvenile polyposis [6]. Polyps are mainly found in the stomach and colon where the carcinogenic risk is higher [7].

Juvenile polyposis is divided to three subtypes: Juvenile Polyposis Coli, JP of infancy and generalized JP. The JP of infancy type is the most severe. It begins in the first months of life with numerous hamartomas of the entire digestive tract responsible for early death by diarrhea, malnutrition or hemorrhage. Macrocephaly and hypotonia are frequently associated. A frequent association with malformations: malrotation, cardiac or genitourinary abnormalities have been reported. In our clinical case, we have a 16 year old patient followed for a congenital coarctation of the aorta and whose diagnosis was revealed by a picture of chronic rectal bleeding [7].

Macroscopically, the polyps are usually spherical, with a smooth surface and a short pedicle. Histological examination of the polyps is necessary for the diagnosis, it shows the presence of an abundant lamina propria with mononuclear cells (macrophages or lymphocytes), without significant arborization of smooth muscle, and presence of cystic glands filled with mucus while the epithelium is generally normal, rarely dysplastic.

Colorectal endoscopic screening is crucial, based on the presence of symptoms in childhood, and systematically in adolescence around 15 years of age, it consists on 1 colonoscopy every 2 years [5]. Endoscopic treatment of polyps is possible in certain less profuse forms (patients with about 20 polyps). Surgery treatment is indicated if the number of polyps is than 20, in case of dysplasia, or if there is a complication. A total colectomy with ileo-rectal anastomosis, or a total coloproctectomy with ileo-anal anastomosis [8] can be performed, which is the case of our patient, a total coloprotectomy was indicated in front of the presence of a colon full of polyps with dysplasia objectified in the rectal polyps.

When it comes to the upper gastro intestinal tract follow-up, the risk of gastric cancer is about 25% during juvenile polyposis and therefore an esophagogastroduodenoscopy every 2 years from 15 years old is recommanded. Same as the colon, gastrectomy is indicated if we find a high number of gastric polyps, complications or dysplasia objectifed at the histological study.

CONCLUSION

Juvenile polyposis is a rare genetic disease which symptoms are not very specific, dominated by rectorrhagia. The diagnosis is essentially endoscopic and histological. Endoscopic screening is essential because of the risk of malignant degeneration of juvenile polyps.

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