Case presentation: villous tumor of rectum in a child

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Abstract

Introduction: The rectum villous tumor is an uncommon pathology in paediatric patients, it represents 1% of all the children's malignant tumors.

Case presentation: An eleven-year-old girl presented a rectal adenocarcinoma arising from a villous tumor. The aim of this study is to overview the literature, asses the frequency of rectal villous tumor, specify the value of the clinical examination, of the radiologic findings in the assessment of the loco-regional extension of villous tumor and rectal adenocarcinoma and finally to discuss the treatment modalities.

Conclusion: The rectal villous tumor symptomatology in children is not very specific. Therefore, a good knowledge of its clinical presentation and the predisposing pathological factors can improve the prognosis of this rare disease.

Keywords

- Paediatrics
- Villous tumor
- Rectal adenocarcinoma
- Surgical treatment

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Introduction

The objective of this study is to assess, through our observation and the literature data, the frequency of rectal villous tumors-especially the histological subtypes, to specify the value of the clinical examination, the endorectal ultrasound scan, the scanner and Magnetic Resonance Imaging (MRI) in the assessment of the loco-regional extension and finally study the therapeutic modalities.

The rectal villous tumors represents 1% of all the children's malign tumors and from 1.4% to 5.4% of the whole recto-colic cancers.[1,2] They are precancerous injuries, deriving from a pre-existing lesion: the adenomatous polyp following an accumulation of complex genetic alterations. Their occurrence should trigger a search for a hereditary predisposition.

They still present specific problems in diagnosis and treatment within the framework of the rectal neoplasic lesions.

These tumors are known for their high potential of malignant degeneration (between 30 and 40 of cases).[3] They are voluminous and web-shaped with a large implantation basis and are more often located in the rectum.[4,5] Their large extension on the surface can furthermore make a radical surgical treatment mutilating, in particular with low rectal lesions.

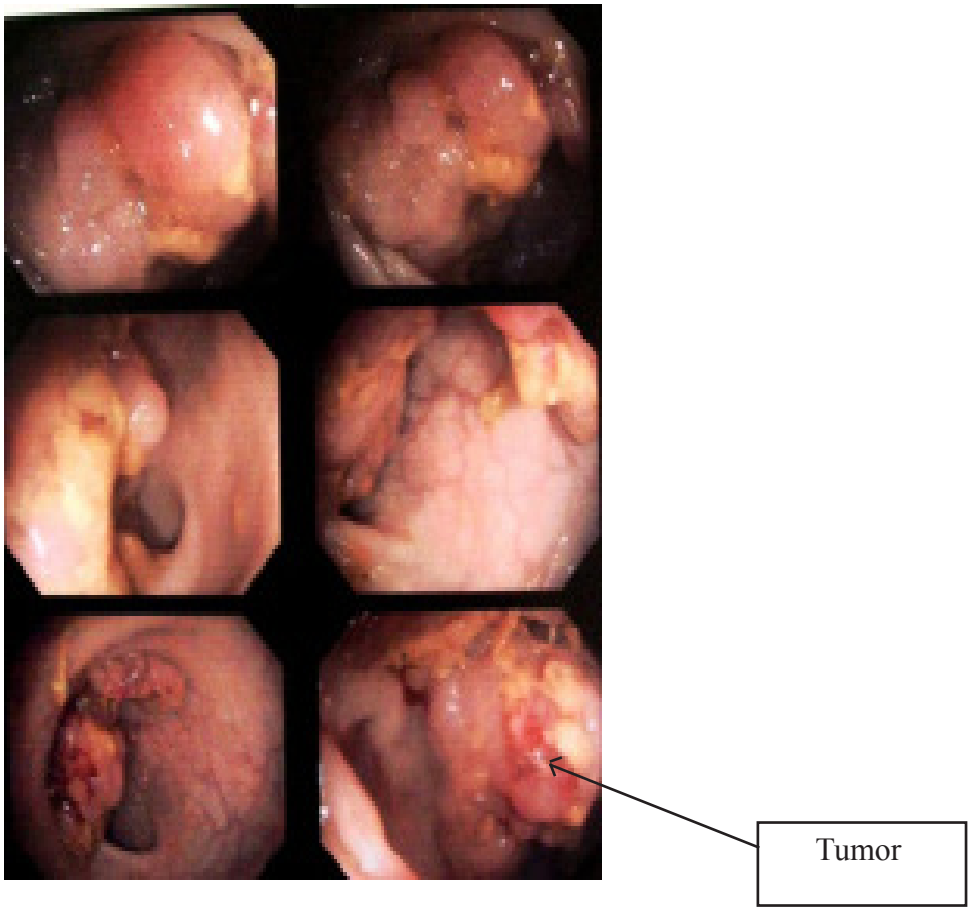
Their poor prognosis can be improved thanks to an early diagnosis, a better knowledge of the ways of presentation and the predisposing pathological situations.

Case presentation

An eleven-year old girl, outcome of a first-degree intermarriage without medical history presented with profuse diarrhea containing phlegm and mucus, accompanied by rectal bleeding, tenesmus and abdominal pain. The symptomatology was resistant to medical treatment.

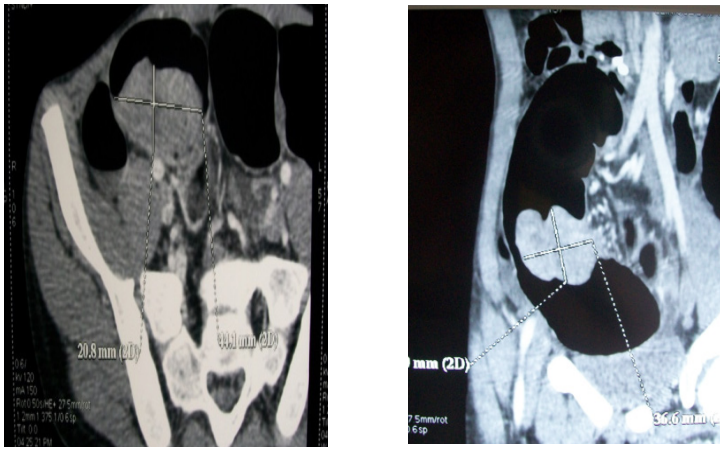
The physical exam showed several light brown spots in trunk and limbs. The abdomen was painful and tender and the rectal examination did not reveal any palpable mass. Colonoscopy revealed a voluminous rectosigmoid polyp of 4.4 cm, sessile, with large implanting, partially obstructive and fragile with other small polyps at the colon **Figure 1**.

Figure 1: Coloscopy: Rectosigmoid polyp, partially obstructive located at 10 cm from anal margin



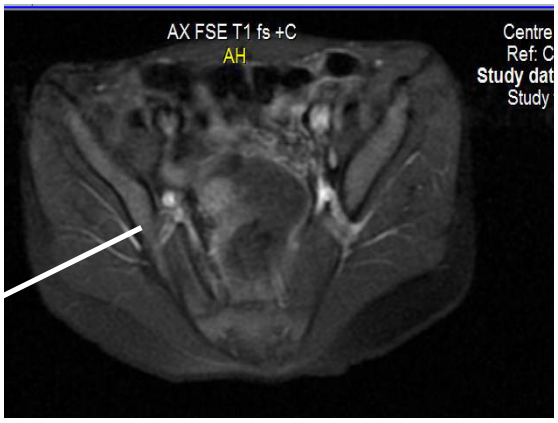
The check-up was completed with a tomodensitometry that showed a polyp with intraluminal development, located within 10cm from the anal margin 44mm from the major axis without evidence of peri colic invasion, together with a mesenteric adenopathy of 14 to 28mm diameter **Figure 2**.

Figure 2: pelvic tomodensitometry axial (A) and sagittal (B) reconstructions with contrast injection polyp tumor located within10 cm from anal margin, with a diameter of 44×33 mm



Abdominal MRI depicted rectocolic polypoid lesions with a discrete high signal intensity T2 weighted and isosignal T1, enhanced after gadolinium injection. Parietal multifocal tumor budding in the rectum and the sigmoid colon was seen **Figure 3.**

Figur 3: Pelvic MRI axial reconstruction T1 weighted with gadolinium injection showed parietal rectal high



Chest radiography and abdominal ultrasonography were normal. The anatomopathological examination showed a villous adenoma with zones of moderate and severe dysplasia, along with of all of an invasive Lieberkuhnian adenocarcinoma crossing the muscularis mucosa and invading the polyp axis.

The invasiveness of the tumor demanded surgical treatment.

First, through a perineal approach (endoanal SOAVE), dissection of the mucosa and submucosa on a 10 cm length was performed.

Then, by laparotomy we found three tumors lying in the sigmoid colon, the descending colon and left splenic flexure **Figure 4**.

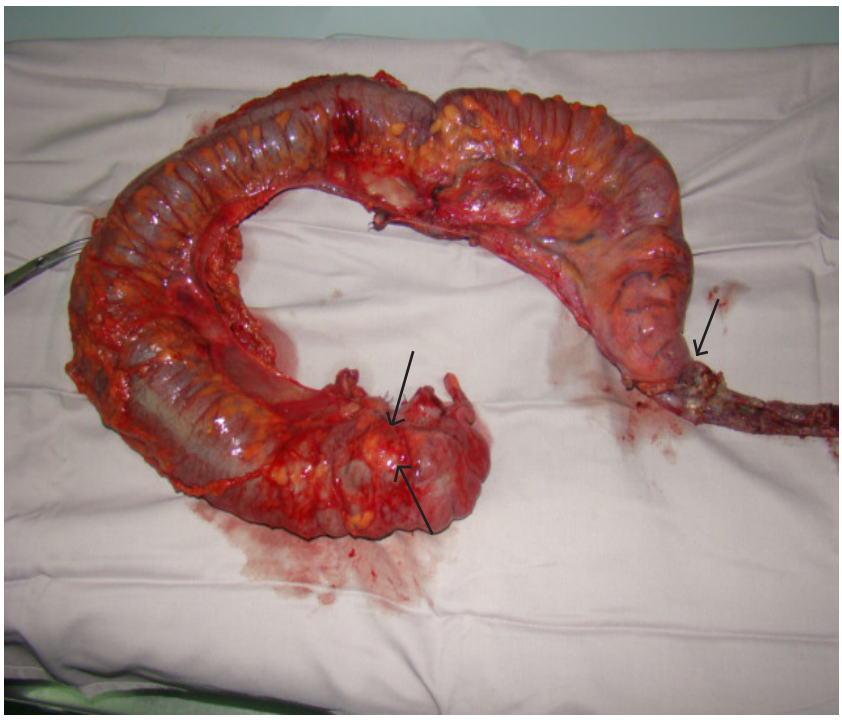
The operation consisted, in a total colectomy with ileoanal anastomosis. Our patient was operated on in June 2010, followed by a chemotherapy treatment.

The diagnosis was confirmed by biopsy. The tumor was classified PT2N1M0.

The post-op chemotherapy was prescribed according to the alternate operating procedure CAMPTO-FUFOL (according to the JCO 2007).

The post-op follow-up was complicated by

Figure 4: surgical specimen: 3 tumors in the sigmoid colon, the descending colon and left splenic flexure (the arrow).



vomiting, dehydration and severe tonic-clonic generalized seizures.

The brain scan was normal, however the abdominal scan revealed an important gastroduodenal distention, a flooding ascites as well as peritoneal carcinosis. Very quickly ionic disturbances settled and this led to death 14 months after the operation.

Discussion

About 500 cases of colorectal cancer have been reported in children. The rectal villous tumors represent 1% of the malignant ones at this early age. The impact before age 20 has been estimated to be around one out of a million cases. It is higher between 10 and 19: 6.8/1million.[1,5]

In our series, the rectum villous tumors represents one case out of 10 000 hospitalizations.

It occurs usually between 9 and 17 years-old with a maximal frequency at the age of fifteen with a slight male predominance.

These tumors are known for their degeneration potential in an adenocarcinoma in 30 to 40% of cases. The rectum represents their predilection area in 95% of the cases.

The earliest genetic anomaly detected in the colorectal cancer is often the APC (Adenomatous Polyposis Cancer) gene mutation which is a suppressor gene located at the level of the chromosome 5 (5q21) long arm locus 21.

These abnormalities were not found in our patient.

The clinical picture is dominated by proctorrhagia as well as large amounts of mucous-glairy depositions with or without abdominal pain.

The colorectal cancer symptomatology is far from being particular to children. The diagnosis is rarely evoked at this age. Most of the authors report a delay in diagnosis with a deadline varying on average between 3 and 6 months, reaching sometimes two years.[1,5,7]

The colonoscopy is essential to the diagnosis. It also allows biopsies and polyp excision.[5]

The rectal endosonography is the most efficient study for evaluation of the the locoregional extension of the rectal cancer. It enables the assessment of the infiltration of the rectal wall, the study of the ganglionic invasion and of the perirectal fat tissue more precisely than by tomodensitometry.[8,9]

It is also useful in the pre-therapeutic assessment of the rectal villous tumors, especially for those situated above and slack at the surface.

The ACE (Antigène Carcino-Embryonnaire) meaning the Carcinoembryonnic Antigen is not a reliable marker of the infant colorectal cancer, contrary to adult.

The ecography, the tomodensitometry, or even the Magnetic Resonance Imagery (MRI) are used for better detecting the metastases that are most often hepatic, pulmonary and vertebral.[5]

The villous adenoma offen appears on

tomodensitometry often as papillary projections reproducing villi, giving a burgeoning invaginated aspect or a cerebral hemisphere or of a cauliflower one, without peri-rectal or pelvic adenopathy. It consists essentially of epithelio-connective digitations hailstorm and branched developing in the intestinal lumen. A malignant component is observed in 40% of cases with a correlation between the size of the tumor and the potential of its malignancy (the risk is six times higher above 2 cm).[1]

Surgery is indicated for tumors less than 4cm height occupying a third of its circumference, tumors located within less than 8cm of the anal margin, and those occupying half of the circumference with a tumorous height between 5 and 6cm.[4,12,13]

This surgical treatment varies according to the lesion size. Most of the tumors are accessible to transanal local excisional surgery while the very extensive lesions require a proctectomy for technical imperatives (high risk of incomplete excision) and oncologic ones (increased risk of degeneration into an invasive cancer).[4-6]

The postoperative treatment is established according to the histological type and the tumor stage: The place of chemotherapy is ill-defined.

The one year survival of a child would not exceed 10%. This prognosis is transformed after surgery.[1]

Conclusion

The child rectum villous tumor symptomatology is not very specific. A late diagnosis is often reported. The prognosis is disastrous, related to the late stages and the adenocarcinoma with a villous component. Its improvement is through increasing the knowledge of modes of presentation and through the mastery of the predisposing pathological situations: polyposis, ulcerous pancolitis and hereditary cancer without polyposis.

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