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Case Report

The Primitive Rectal Plastic Linitis: Role of the Endoscopist

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Abstract

Primary rectal linitis is a rare and aggressive tumor; of young people. Because of its invasive potential; the diagnosis is often discovered late. We report the case of a 16y.o patient admitted for a six-month-old rectal syndrome and severe weight loss. The evolution and prognosis were poor after 8 months. The role of endoscopy was to get deep specimen for pathological diagnosis to avoid surgical excision.

Keywords: *Linitis; Rectum; Rectoscopy*

Introduction

Primary rectal linitis is a rare tumor; but extremely aggressive; which mainly affects young people. Because of its invasive potential; the diagnosis is often discovered late.

Case Report

A 16-year-old patient was admitted for a six-month-old rectal syndrome in the context of severe weight loss. The abdominal examination was normal. The rectal examination perceived at 3 cm of the anus a painless hard circumferential mass. The other clinical examination was normal. At 3 cm from the anal margin, rectoscopy revealed a circumferentially nippled tumor process (Figure 1A-1D). The biopsy revealed infiltrating carcinoma of the rectum as a "pintle ring" (Figure 2). The abdominopelvic computed tomography showed a recto-sigmoid wall thickening of tissular density enhancing after contrast injection with extension to perianal fat and adjacent lymphadenopathy. The search for a primary localization consisting of a high endoscopy, a vesicoprostatic ultrasound with a PSA and liver functional data was negative. The barium opacification showed a rectal stenosis. The diagnosis of primary rectal linitis was retained and surgical exploration found a rectal lesion process invading the sigmoid loop, in relation to the posterior plane with carcinomatosis, lymph nodes along the aorta and ascites. The patient had received two courses of chemotherapy combined with radiotherapy. The evolution had been marked by a significant alteration of the general condition. The patient died 8 months later.

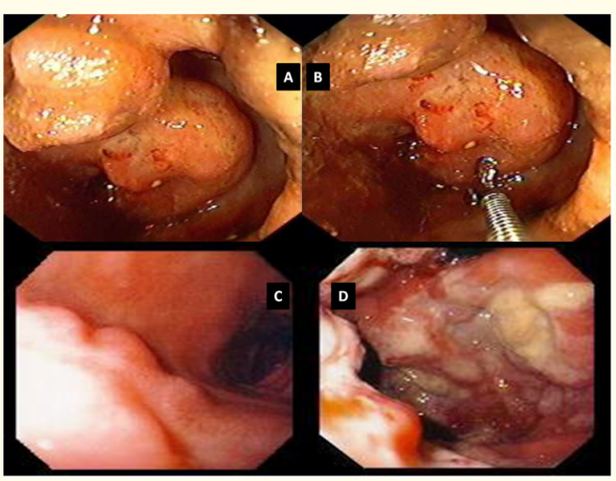

Figure 1: *Endoscopic view of a primary rectal linitis (A, C, D: budding aspect, B: biopsy with the forceps).*

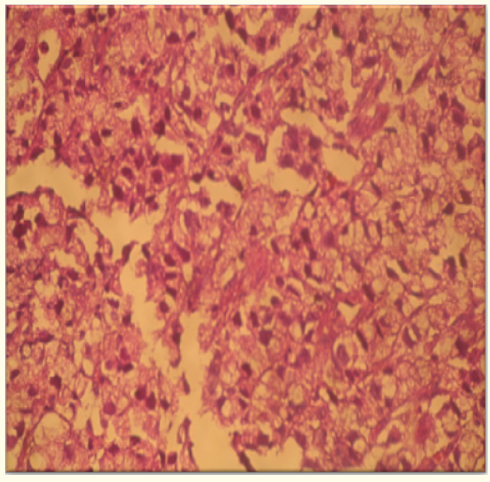

Figure 2: *Pathological aspect: tumor cells in a kitten ring (PAS staining).*

Discussion

Primary rectal linitis is a very rare pathology. According to the series it represents 0.2 to 0.4% of rectal cancers [1]. Fahl., *et al.* [2] reports an incidence of less than 1/1000 for a series of 12000 colon cancer. However, in Jordan and Lebanon, rectal linitis accounts for more than 18% of colorectal cancers [3,4]. There is a discreet male predominance with a sex ratio of 1.3 [5,6]. The average age varies in the literature between 52 and 67 years [7,8] it is lower than that of colorectal adenocarcinoma [9,10]. The average age of our patients is lower than the average age of our colorectal cancer cases, which is 50 years old. It is obviously a primitive limite since no other localization is evidenced in our patient and upper digestive fibroscopy has cleared the stomach, the organ of choice of this affection.

The symptomatology of the rectal linitis is not specific. It can be revealed by transit disorders (76%), abdominal pain (60%), weight loss (40%) [11]. However, it is characterized by the absence of bleeding because the mucosa is not ulcerated [12]. The rectal examination often specifies the stenosing and infiltrating character of the lesion [13,14].

In the barium opacification, the linitis is in the form of a long, regular and centered stenosis, without ulceration or intraluminal process, with a rigid aspect of the wall. It often evokes inflammatory stenosis [14]. At endoscopic exploration, the lumen is narrowed, sometimes responsible for an insurmountable stenosis, the mucosa is intact. Biopsies do not reveal tumor lesions in 50% of cases [11]. This why the endoscopist should be able to have a deep and controlled specimen to not perforate the digestive wall. Indeed tumor cells sit at the level of the submucosa and spare the mucosa, biopsies must be deep. In our patient, several biopsies were negative including three deep biopsies under general anesthesia. False reassuring results from deep biopsies have been the cause of delayed diagnosis and non-resectability of the tumor. CT and MRI show stenosis and thickening of the rectal wall. They make it possible to evaluate loco-regional invasion. Echo-endoscopy shows hypoechogenic circumferential thickening reaching the submucosa and the muscularis mucosa. This examination is limited by the existence of a stenosis, but it remains the most effective for the diagnosis of small tumors and lymph node metastases [13].

The diagnosis of certainty of rectal linitis is revealed by the histological study of deep biopsies performed endoscopically or surgically, or by the study of the surgical excision specimen. The typical macroscopic appearance is characterized by a concentric and rigid thickening of the intestinal wall extending over several centimeters [11].

Histologically, linitis is defined by two criteria. The first is the presence of more than 50% of independent cells in "kitten ring" [11]. These cells are characterized by a big vacuo the mucus that drives the nucleus to the periphery of the cytoplasm. The second criterion is the inclusion of these cells in a stroma that is fibrous, dense and abundant. These elements infiltrate the entire wall respecting the architecture of the layers. The mucosa is intact. The search for a primitive extra-rectal localization, especially gastric, by echo-endoscopy with biopsy is imperative.

The differential diagnosis is with radiation and ischemic colitis, lymphomas, endometriosis, tuberculosis, pelvic-rectal inflammatory pseudo tumors, but the confusion is mainly with inflammatory bowel diseases, especially since the association of the two diseases is possible. In the literature, six cases of colorectal plasticitis were associated with ulcerative colitis and one with Crohn's disease [15]. The role of a genetic, racial predisposition or the degeneration of an adenomatous polyp has not been established. The treatment of plastic linitis joins that of rectal cancer. It combines carcinologic excision of the rectum and lymph node dissection. Preoperative radiation therapy, combined with 5-Fluoro-Uracil and cisplatin-based chemotherapy, appears to improve survival [16]. The prognosis of this condition is severe, due in part to the existence of lymph node extension (86%), pelvic (58%), peritoneal (47%). Hepatic metastases are rare [5]. The average survival of the patients according to the literature is 9 months after the diagnosis [16]. Our patient (observation 1) survived 19 months after the appearance of the first symptoms, without any radical treatment while our second patient and after receiving two courses of chemotherapy combined with radiotherapy. The evolution was marked by a significant deterioration of the general state and a death occurring 8 months later.

Conclusion

The primary linitis of the rectum is a rare form of rectal cancer diagnosed most often at a late stage, which makes the prognosis very pejorative. The diagnosis of certainty is mainly histological, obtained on deep biopsies.

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