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# Primary Anorectal Melanoma: A Rare and Aggressive Location

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**Abstract** **Case Report**

The primary anorectal melanoma is an aggressive tumor with poor prognosis; its diagnosis is often delayed because of the scarcity of the entity and confusion with other benign anorectal lesions. The management consists of complete surgical resection, however the majority of patients are diagnosed at the metastatic stage. We report the case of a 52-year-old man who consulted for chronic anal pain, proctologic examination for an anal tumor, and histopathological analysis with immunohistochemistry for anorectal melanoma. The patient started chemotherapy sessions as the extension assessment showed liver metastases.

**Keywords:** Anorectal melanoma, prognosis, chemotherapy.



## INTRODUCTION

The anorectal melanoma is a rare and aggressive tumor. It accounts for about 1% of all [1, 2] melanomas and about 0.5 to 2% of all anorectal malignancies [3, 4]. It is the most common location of melanoma after the skin and eyes, it occurs most frequently in women [5, 6], its diagnosis is often difficult as it does not have specific symptoms. Rectal bleeding, the most frequent symptom, is present in 55% of cases [7]. More rarely, the patient reports: pruritus, tenesmus, transit disorders or prolapsed hemorrhoid [8]. Tumor extension is often at the level of the submucosal planes explaining the importance of loco regional infiltration and the frequency of metastases at the time of diagnosis [9]. The prognosis is poor, with a median survival of 24 months to five years for 10-15% of patients [10]. Early diagnosis is essential to improve the survival rate of these patients. Conventionally, treatment consists of complete surgical resection [11], the majority of the patients progress to metastatic disease and the use of chemotherapy has been advocated in such cases to improve overall survival [12]. We report a case of anal melanoma at the metastatic stage.

## CASE REPORT

This is a 52-year-old patient with no particular medical condition history including no smoking, no co-morbidities, negative HIV serology, no immunosuppression, no history of melanoma lesion excision, with no other individual or familial medical condition history, who has consulted for chronic anal pain with a few episodes of low-abundance rectal bleeding without transit disorders and without other associated digestive or extra digestive symptoms.

The physical assessment found a man with impairment of general condition, BMI was 17 kg / m2, WHO score 2, no findings on abdominal examination and anal margin examination. On digital rectal examination: perception at the level of the anterior wall and as early as 1 cm of anal margin, a fixed and painless induration with preservation of sphincter function, no palpable peripheral inguinal or supraclavicular peripheral lymph nod and the remainder of the clinical examination, especially cutaneous and ocular was normal. Recto sigmoidoscopy revealed at the level of the anterior wall at 01 cm of anal margin, a circumferential budding tumor process bleeding in extensive contact about 4 cm (biopsies done) without internal hemorrhoids (Fig-1).

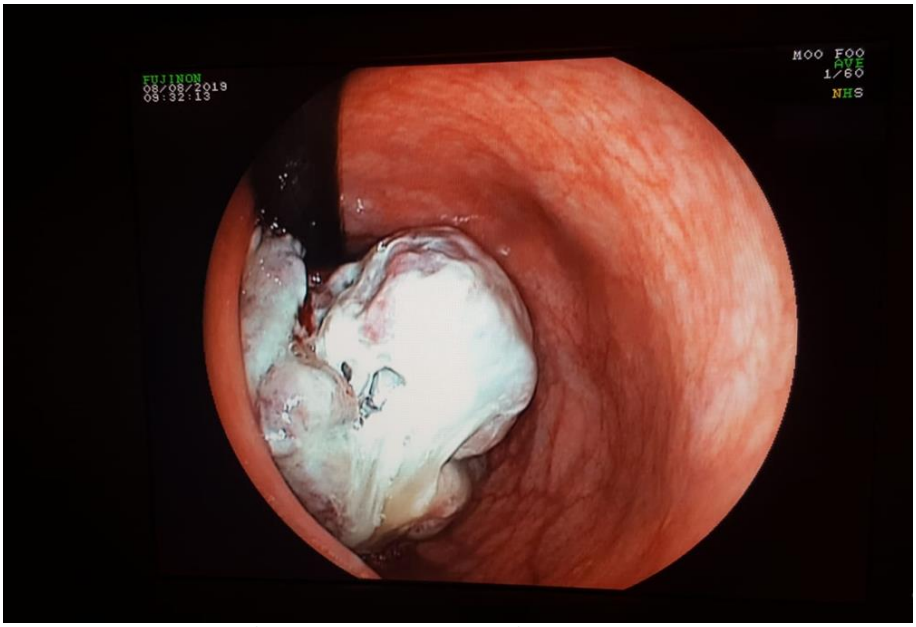


**Fig-1: Rectosigmoidoscopy showed at the level of the wall anterior to 01 cm of the anal margin, a budding tumor process extended about 4 cm without internal hemorrhoids**

On histopathological examination with immunohistochemical study, the tumor cells were highly positive for HMB-45 and Melan A, thus the diagnosis of melanoma was retained.

As part of the extension assessment, a thoracoabdominal and pelvic CT scan was performed, which showed a thickening circumferential tumor process of the extended rectum towards the anal canal with liver metastases. Pelvic MRI confirmed the results of CT (Fig-2) with evidence of circumferential tumor thickening of the low rectum extended to the anal canal measuring 28 mm in thickness, extended to 43 mm in height and whose pole inferior comes into contact with the anal margin with respect to the sphincter apparatus and neighborhood structures with large pelvic lymph nod over the bladder measuring 34 * 36 mm and prostatic hypertrophy. Thus the tumor was classified metastatic stage IV (T2N1M1) and the patient started the chemotherapy sessions.

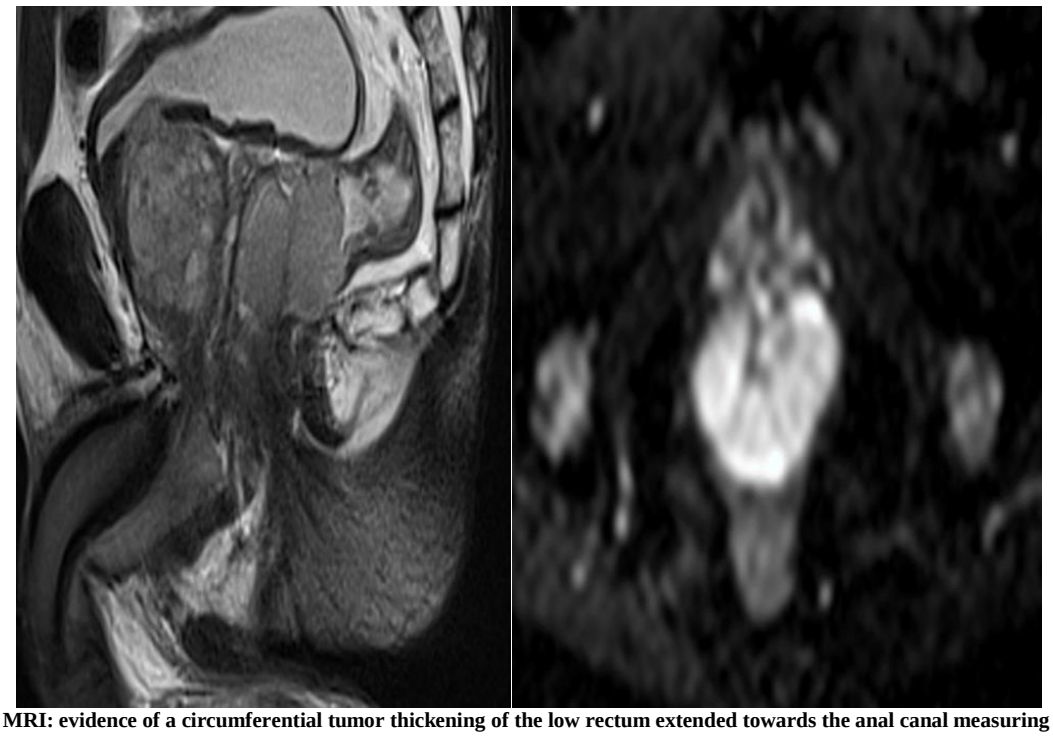

**Fig-2: Pelvic MRI: evidence of a circumferential tumor thickening of the low rectum extended towards the anal canal measuring 28 mm thick, spread over 43 mm in height and whose lower pole comes into contact with the anal margin with Respect of the sphincter apparatus and neighborhood structures with voluminous pelvic adenopathy over the bladder measuring 34 * 36 mm and prostatic hypertrophy**

## DISCUSSION

The melanoma is a tumor that develops from melanocytes derived from the neural crest. During the fetal life, these cells migrate to several sites of the body such as the skin, the retina and the ano-rectal mucosa [4]. The rectum has a typically glandular epithelium. In contrast, the anal canal below the pectine line is covered with squamous epithelium. Above the dentate line is the transition zone where the glandular and squamous cells are present. Melanocytes may appear in all three regions (rectum, anal canal and transition zone), although the presence of melanoma is more common in the transition zone and squamous epithelium [13]. The clinical symptoms of anorectal melanoma are mainly: rectal bleeding, anal pain and perianal mass. Because of the non-specificity of the clinical picture, 60% of patients are already at the stage of metastasis at the time of diagnosis [14]. Proctological examination with inspection, digital rectal examination and rectoscopy is extremely important because it provides data such as size, consistency, fixation and invasion of the sphincter or adjacent structures [15].

The ano-rectal melanoma is often a polyploid or nodular lesion of blackish or brownish color. But in 30%, it can be amelanotic [16] as for our patient, the macroscopic appearance can mimic that of benign lesions such as thrombosed hemorrhoids or a polyp [17-19]. The histological diagnosis is based on the detection of melanin pigment in the tumor cells. In the amelanotic forms, the immunohistochemical study is essential, it makes it possible to objectify a positive immunolabeling for the protein S-100, vimentin and the specific antibodies of the melanoma: HMB-45 and Mart-1 [16]. In our case, the immunohistochemical analysis was positive for the protein HMB-45 and Melan A.

The primitive character of ano-rectal melanoma should be retained only after careful questioning and clinical examination given the frequency of digestive metastatic localizations of melanomas [20, 21].

Ano-rectal melanoma can appear in three forms: limited to the primary site, disseminated to regional lymph nodes or with distant metastases. The diffusion can be local or by lymphatic or hematogenous pathways. In lymphatic dissemination, the lymph nodes most commonly affected are those of the inguinal, obturator, mesenteric and para-aortic chains [22]. In hematogenous diffusion, the most frequent sites are: liver, lungs, brain and bones.

The prognostic factors include the stage of the disease at the time of diagnosis and the thickness of the tumor [23].

Due to the scarcity of ano-rectal melanoma, its optimal treatment remains controversial. Complete local resection is currently the therapeutic option of choice for operable patients. It allows disease control with a better quality of life compared to abdominopelvic amputation [24] and a comparable survival [25]. But at the time of diagnosis, 25% of patients are considered no operable because of the presence of metastases or the importance of locoregional extension [26] as in our case. Chemotherapy has not proven effective in the treatment of ano-rectal melanoma. Several molecules have been used, such as dacarbazine, nimustine and cisplatin, but without any significant impact on survival [27, 28]. The role of radiotherapy is also limited. The prognosis of ano-rectal melanoma remains poor, the overall survival at five years varies between 16 and 34% [29]. The average survival is 34 months for localized tumors and 10 months for metastases [11].

Kim *et al*., [30] performed a retrospective review of 18 patients with metastatic anorectal melanoma treated with cisplatin-based chemotherapy in combination with interferon alpha-2b or interleukin-2. They reported that combination chemotherapy was effective against metastatic ano-rectal melanoma.

## CONCLUSION

Our observation emphasizes the importance of early detection of anal melanoma and exposes the difficulties of therapeutic management in the absence of established recommendations. Currently, the new targeted medical treatments and immunotherapies open new perspectives of treatment in patients with anorectal melanoma.

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