

Case Report,

Primitive Anorectal Malignant Melanoma: A Rare and Aggressive Localization a Case Report

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Abstract:

Primary anorectal malignant melanoma is an extremely rare condition. It appears at the third highest frequency after melanomas of the skin and retina. Its prognosis is dreadful because of the early onset of metastases. The treatment remains essentially surgical. We report an observation of primitive anorectal melanoma, collected at the department of surgery for digestive cancers and liver transplantation of the Ibn Rochd University Hospital of Casablanca, with a review of the literature. In order to analyse the clinical, paraclinical and therapeutic characteristics of primary anorectal melanoma.

Key words: melanoma, rectum, abdominoperineal resection, prognosis.

Introduction:

Malignant melanomas are malignant tumors that develop at the expense of the pigment system [1]. Primary localization remains a very rare entity, less than 2% of melanomas. The corner stone of treatment remains surgery. The prognosis is bleak as the diagnosis is often made at advanced stages and their metastatic potential is high [2].

Observation:

The patient was 68 years old, with no particular pathological history, and had presented 6 months before admission with low abundance rectorragies, a rectal syndrome and proctalgia; all of which evolved in a context of weight loss of 10 kg in 6 months and an alteration of the general state. The clinical examination at admission had objective a Performance Status (PS) of 1 and a Body Mass Index (BMI) of 22 kg/m². The abdominal examination was unremarkable, and on rectal examination there was a hard pedicled mass prolapsed by the anus at the expense of the anterior wall of the anal canal a 1cm from the anal margin

extending over 4 cm (figure 1). The anatomopathological examination of the biopsy carried out had revealed an undifferentiated ulceronecrotic large-cell undifferentiated malignant tumor proliferation, which had been pigmented focally, and whose complementary immunohistochemistry showed a positive antibody response to Melan A, Ki67 and HMB 45, thus indicating an infiltrating large-cell undifferentiated tumor proliferation compatible with a rectal malignant melanoma. Colonoscopy had not revealed any other lesions. The CT scan showed a thickening of the lower rectum with normal perirectal fat with no other locoregional or secondary localizations. Pelvic MRI (Figure 2) showed a thickening of the lower and middle rectum that could be classified as T3N1Mx with sphincter infiltration and significant endometrial thickening. The 18-fluorodeoxyglucose PET-CT scan showed hypermetabolic process of the anal canal and lower rectum suvmax10.9 metabolic volume 28.3 cm³. The file was staffed in a

multidisciplinary consultation meeting and the decision was to perform an abdominal perineal amputation (AAP) with hysterectomy. The surgical treatment consisted of an extra-levatorial AAP with hysterectomy (figure 3) and a definitive iliac colostomy. The postoperative follow-up was simple and the patient was declared discharged on day 4 postoperatively. Histological examination of the operating specimen revealed malignant tumour proliferation of the round-cell rectum compatible with melanoma and an endometrial polyp in the uterus without signs of malignancy. The immunohistochemical complement showed tumour proliferation expressing PS100 and Melan A but not expressing CKAE1 or AE 3 or Ckit or chromogranin or CD 45. Thus, the diagnosis of achromic malignant melanoma of the rectum was retained with a tumor thickness of 2cm and 15 out of 15 lymphnodes were negative in the mesorectum. The setback is one year.

Discussion:

Primary anorectal melanoma is a rare tumour first described in 1857 by Moore. It accounts for 0.1-0.5% of anorectal cancers and 1.5% of all melanomas. It appears third in frequency after melanomas of the skin and retina. The average of onset is around 50 years with equal frequency in both sexes [3]. From an etiopathogenic point of view, the most probable hypothesis is that of chronic irritation, given the exclusion of sun exposure in this location. The diagnosis of primary anorectal malignant melanoma can only be made in the absence of any synchronous localization (skin, retina), and the absence of a history of melanoma excision, whatever its location, which was the case in our patient [4]. Clinically, it is mainly manifested by rectorragies, anal pain, perianal mass, rectal syndrome and/or inguinal mass related to inguinal adenopathy [2]. Due to the non-specificity of the symptomatology, approximately 70% of patients are diagnosed with advanced metastases, which may contribute to the high recurrence rate and poor survival results [5, 6]. On proctologic examination, primary anorectal melanoma is almost often presents as an ulcerative vegetal tumor or a polypoid, pedicled lesion. The characteristic black colour of melanoma is present in our patient [4]. The diagnosis is histological based on the detection of melanin pigment within the tumourcells. In the amelanic forms, immunohistochemical study is essential, making it possible to demonstrate a positive immunostaining for the protein S-100, vimentin and melanoma specific antibodies: HMB-45 [7]. Anal echo-endoscopy

makes it possible to evaluate the extension of the melanoma in the rectal wall as well as the perirectal lymphnode involvement. The assessment of locoregional and remote extension is essentially based on CT scan, allowing detection of pelvic lymphnodes and visceral and bony metastases. It is an important tool for the follow-up of treated patients [6]. Several classifications of primary anorectal melanoma have been proposed. The Slingluff classification classifies melanoma into 3 progressive stages. It is of prognostic interest [8]. In the absence of clear consensus due to the lack of randomised studies, the overall objective of treatment must be to optimise quality of life and tumor control while minimising treatment-related morbidity [4,9]. Therapeutic options are represented primarily by surgery, most often performing AAP with inguinal and pelvic lymphnode dissection, which appears to be a major prognostic factor influencing survival, or localized tumor resection, the use of one of the methods remains controversial [4]. A Japanese meta-analysis is conducted in 2015 by Matsuda et al. [10] concluded that AAP has no benefit in terms of patient survival. Since local failures after local resection could be managed by salvage surgery, prophylactic lymphnode dissection is indicated only if pelvic adenopathy is present [10]. Other therapeutic means such as chemotherapy and radiotherapy are currently used as palliative treatment because their efficiency in the curative management of melanoma remains to be approved. Studies are currently converging towards immunotherapy, which also has not been proven to be effective [11, 12].

Conclusion:

Anorectal melanoma remains a challenge. Efforts must be made for early diagnosis. Treatment remains essentially surgical and wide local excision with healthy margins is the preferred treatment. AAP is a reasonable option for large tumors or when the sphincter is invaded. Targeted therapy and immunotherapy are promising therapeutic means that can improve the prognosis of anorectal melanoma.

Conflict of Interest:

The authors do not declare any conflict with interest.

Authors' contribution:

All authors also contributed to this work and read and approved the final version of the manuscript.

Figures:

Figure 1: Intraoperative view of the tumor prolapsed by the anus (black arrow).

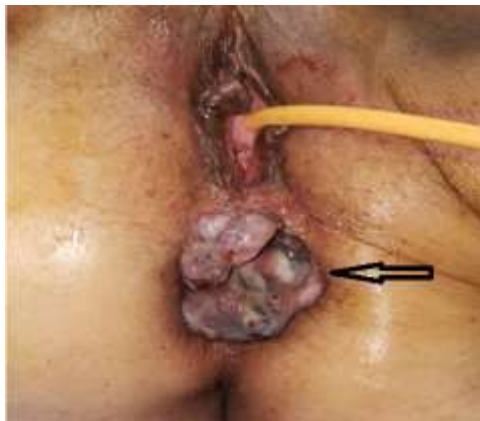


Figure 1: Intraoperative view of the tumor prolapsed By the anus (black arrow).

Figure 2: Sagittal section of pelvic MRI: Showing a thickening of the lower and middle rectum (yellow arrow).

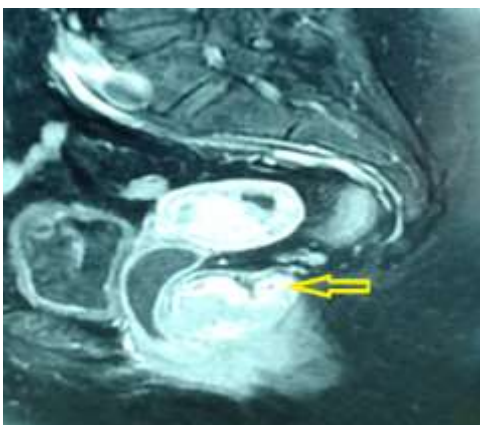


Figure 2: Sagittal section of pelvic MRI: showing a thickening of the lower and middle rectum (yellow arrow).

Figure 3: Operating specimen of a monobloc resection of abdominal perineal amputation (yellow star) with hysterectomy (black star).



Figure 3: Operating specimen of a monobloc resection of abdominal perineal amputation (yellow star) with hysterectomy (black star).

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