IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 22, Issue 7 Ser.12 (July. 2023), PP 05-07
www.iosrjournals.org

A Rare Case Of Malignant Anorectal Melanoma

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ABSTARCT
Anorectal melanoma is rare but aggressive malignancy that poses a significant challenge to physicians due to its non-specific presentation and often late diagnosis.
The authors present a case of anorectal malignant melanoma. This is a rare condition with a poor prognosis. Currently, surgical management is the only therapeutic approach available.
Key Word: *Anorectal melanoma; Aggressive tumor; Endoscopy*

Date of Submission: 21-07-2023 Date of Acceptance: 31-07-2023

I. INTRODUCTION

Anorectal melanoma (ARM) is a rare and aggressive tumor. About 1% of all anorectal carcinomas are melanomas and approximately 0.5 to 2 % in all anorectal malignancies. The anorectal localization is the third after the cutaneous and ocular localization. It usually occurs in the 50s or 60s of life and primarily affects women.
The most commonly reported symptoms include anal bleeding, abdominal pain and a changed defecation pattern.

The prognosis is very poor, with an average survival of 24 months and a 5-year survival rate of 10%. Almost all patients die of metastasis.

Due to the rarity of this disorder, there is no consensus as to which surgical approach is beneficial. The surgical options of choice range from an abdominoperineal resection (APR) to wide local excision (WLE) with or without adjuvant radiotherapy.

II. CASE PRESENTATION

We report a case of ARM in a 69-year-old man with a history of chronic obstructive pulmonary disease, presenting a specific clinical symptomatology, with episodic perineal pain, and rectal bleeding evolving for 4 months. Examination reveals conjunctival pallor secondary to anemia at 9 g/dl.

The proctologic examination associated a rectoscopy reveals a huge ulcero-budding tumor of stony consistency, hemi-circumferential non-stenosing bleeding spontaneously, measuring 3 cm from the anal margin and extended over 3 cm.

The histopathology of the biopsy concluded to a malignant melanoma based on the tumor profile on immunohistochemistry: HMB45+, Melan A +, Wide spectrum cytokeratins negative. (Figure 1)

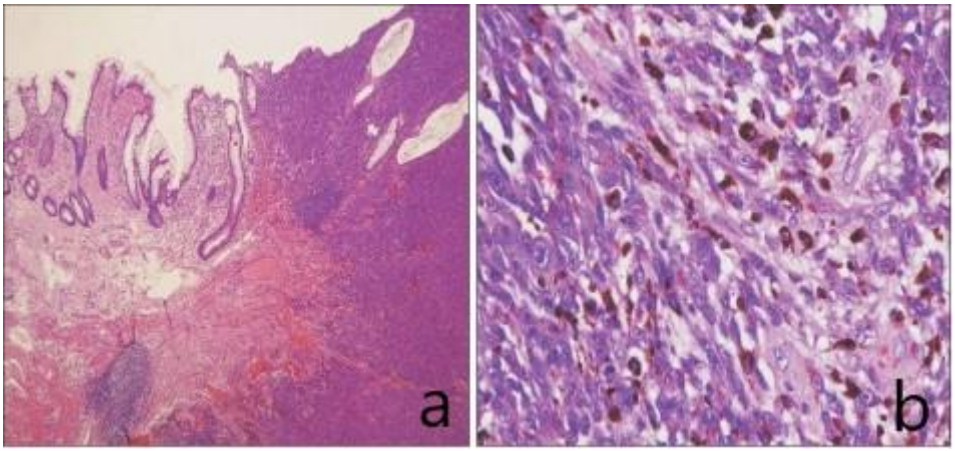

Fig 1. a) microscopic findings revealed diffuse infiltration of round or spindle-shaped tumor cells, and the contained melanin and tumor cells were immunohistochemically (b) positive for Human Melanin Black-45 (HMB-45).

The thoraco-abdomino-pelvic scanner in favor of a tumoral process of the lower rectum and didn't show any evidence of locoregional or distant metastasis.

Magnetic resonance imaging of the pelvis highlighted an infiltration of the upper extremity of the internal sphincter and right obturator adenopathy (Figure 2).

The patient is then referred to the surgical department for abdominoperineal amputation.

The patient died 3 weeks after surgery following an exacerbation of chronic obstructive pulmonary disease.

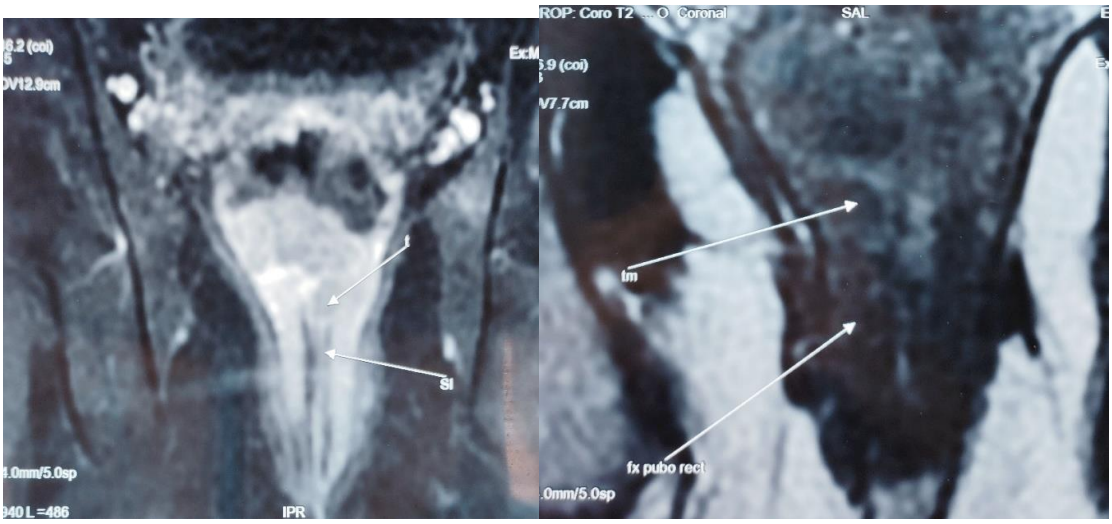

Fig 2. Pelvic MRI: frontal view, showing a rectal tumor (t) infiltrating the internal sphincter.

III. DISCUSSION

Anorectal melanoma is rare but aggressive malignancy that poses a significant challenge to physicians due to its non-specific presentation and often late diagnosis [1]. This tumor commonly affects women in their 50s and 60s with a 1.7-fold higher prevalence in caucasians than in African Americans [2]. It has a low incidence, accounting for less than 1% of all melanoma and less than 4% of all anorectal cancers [3]. However, mortality is high, with a 5-year survival rate of less than 10% [4].

The clinical symptomatology is varied and not non-specific dominated by rectal bleeding, proctalgia and rectal syndrome [5], Our patient had a similar presentation. On proctologic examination, anorectal melanoma often presents as an ulcerobudding tumor or a polypoid lesion. The characteristic blackish color of melanoma is present in a third of cases [6]

Histological confirmation is based on the detection of melanin pigments within the tumor by conventional Fontana staining [7]. The use of immunohistochemistry panels, including S-100 protein, Melan A, HMB 45 and tyrosinase is helpful in diagnosis [8]. In our case, immunohistochemistry analysis was positive for the HMB 45 and Melan A.

Prognostic factors include disease stage at diagnosis [9] and tumor thickness [10]. The most common sites of distant metastasis are the liver and lungs.

The main treatment option for anorectal malignant melanoma is surgery, The extent of resection depends on the stage of the disease, but local excision and abdominoperineal resection are usually the preferred surgical options [11].

Initially, abdominoperineal amputation was considered the technique of choice for local control of the disease, However, several recent studies suggest that, if possible, sphincter-sparing local excision and adjuvant radiation is well tolerated and can effectively control loco-regional disease while avoiding the functional morbidity of the APR [12].

If an APR is performed, the mesorectal lymph node resection may contribute to a better staging of the disease. There is no value of prophylactic inguinal lymph node resection [13].

Patients without lymph node metastasis have a survival advantage with a 5-year survival rate of 20 versus 0% in patients with metastasis. For our patient the treatment of choice was abdominoperineal amputation [14].

For local forms, endoscopic treatment with submucosal dissection has recently been proposed as an alternative treatment to surgery [15].

Radiotherapy, chemotherapy, and immunotherapy are also used to treat anorectal melanoma as an adjuvant palliative treatment to surgery do not seem to bring a gain in survival compared to surgery alone. Radiation therapy has reported to provide a better local control after WLE and also seems beneficial for sphincter preservation [16].

Metastatic disease is associated with a poor prognosis, and data on effective systemic therapy for this rare melanoma subtype are lacking. Currently, standard systemic therapy for cutaneous melanoma is the main treatment for metastatic anorectal melanoma [17].

Kim et al performed a retrospective study 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 for metastatic anorectal melanoma. Responses were similar to cutaneous melanoma [18].

IV. CONCLUSION

ARM is a rare and aggressive disease with a low survival rate. That poses a significant challenge to clinicians due to its non specific presentation and often-delayed diagnosis

Although the optimal treatment has not yet been established, surgery remains the only effective therapy. Further research is needed to develop therapeutic strategies tailored to this malignancy. Continued research on early diagnosis, multidisciplinary approaches, and more effective treatments is essential to improve outcomes for patients with anorectal melanoma.

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