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Anorectal Malignant Melanoma Post- Hemorrhoidectomy

Melanoma Maligno Anorretal Após Hemorroidectomia

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ABSTRACT

Anorectal malignant melanoma, a rare and aggressive tumor, lacks specific symptoms and frequently presents diagnostic challenges due to its similarity to benign anorectal conditions. This case report describes the diagnostic and treatment process of incidentally discovered anorectal malignant melanoma post-hemorrhoidectomy, guided by the existing literature.
Keywords: Anus Neoplasms; Melanoma; Rectal Neoplasms

RESUMO

O melanoma maligno anorretal, um tumor raro e agressivo, carece de sintomas específicos e apresenta frequentemente desafios diagnósticos devido à sua semelhança com doenças do ânus e do recto benignas. Este relato de caso descreve o processo de diagnóstico e tratamento de um melanoma maligno anorretal descoberto incidentalmente após hemorroidectomia, e que foi orientado pela literatura existente.
Palavras-chave: Melanoma; Neoplasias do Ânus; Neoplasias Retais

INTRODUCTION

Anorectal malignant melanomas are rare but highly aggressive tumors, posing diagnostic challenges due to their nonspecific clinical presentation. Unfortunately, the attribution of nonspecific symptoms to benign anorectal conditions often leads to delayed diagnosis, and the disease is frequently incidentally discovered.[1,2] Approximately 30% to 34% of cases present as amelanotic lesions, resembling benign polyps.[1,2] This report details a case of anorectal malignant melanoma diagnosed post-hemorrhoidectomy based on symptoms and appearance associated with hemorrhoidal disease.

CASE DESCRIPTION

A 59-year-old female patient presented with intermittent rectal bleeding, anal soiling, and prolapse of hemorrhoids symptoms persisting for about a year. The patient, diagnosed with hemorrhoidal disease at another center, adhered to lifestyle modifications, including sitz baths, increased fluid and fiber intake, constipation relief, and avoidance of straining. Additionally, she received treatment with micronized purified flavonoid fraction tablets, along with topical applications containing tribenoside and lidocaine hydrochloride. Upon proctological examination of the patient who presented to another center due to the failure of conservative treatment, it was reported that a prolapsed hemorrhoidal plexus, located just proximal to the dentate line, with a relatively smooth surface and bulging lumen, was observed (Fig. 1). Subsequently, an open hemorrhoidectomy was performed on the patient at this center.

On postoperative day 12, the patient presented to our hospital upon suspicion of malignant melanoma based on the hemorrhoidectomy specimen. During the proctological examination performed in the Jack-Knife position, a surgical ulceration was identified in the anal canal. The ulceration began distally from the dentate line at the 7 o'clock position, measuring approximately 4 x 2 cm, and extended proximally towards the rectum.

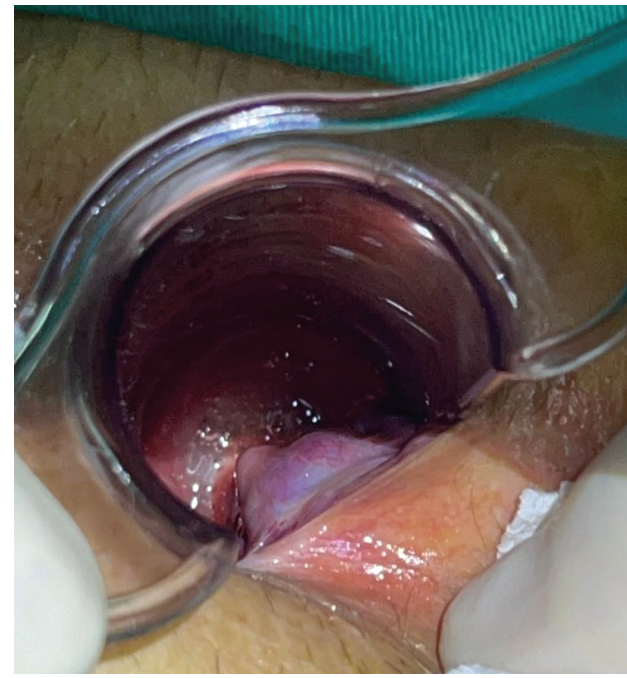

Figure 1 – The appearance of the lesion before hemorrhoidectomy

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Pathological reevaluation at our center confirmed the diagnosis of mucosal malignant melanoma, with the presence of nine mitoses at 5x magnification and tumor infiltration into the rectum. Immunohistochemistry revealed diffuse strong positivity for HMB-45 and Melan-A (Fig. 2). Colonoscopy revealed no pathological findings, while pelvic magnetic resonance ımaging (MRI) identified an ulcerative-defective area extending into the intersphincteric space at the 1 - 2 o'clock position.

Several round-shaped lymphadenopathies, the largest measuring 7 mm, were observed in the perirectal area (Fig. 3). Fluorodeoxyglucose positron emission tomography (FDG PET)/MRI imaging showed increased pathological 18F-FDG uptake (SUVmax: 12.9) in the anal canal and increased pathological uptake in the left presacral and left perirectal regions, with the largest lymph node measuring 9 mm in diameter (SUVmax: 4.2) (Fig. 3).

The case was discussed in a preoperative multidisciplinary tumor board. The patient underwent laparoscopic abdominoperineal resection (APR) and was discharged on postoperative day three. The pathological examination confirmed that the case was a mucosal melanoma and revealed no residual tumor, but two out of 29 excised lymph nodes showed melanoma metastasis. No BRAF mutation (V600E, V600K, V600D, and V600R) was detected.

The patient, for whom systemic treatment was not recommended by the Medical Oncology Department, has been placed under close monitoring. At the six-month follow-up, the patient shows no clinical or radiological evidence of metastasis or locoregional recurrence.

DISCUSSION

Anorectal melanomas represent less than 1% of all melanomas but comprise approximately 4% of anal malignancies.[2] These highly aggressive tumors have a five-year survival rate of less than 10%, with an average survival of 24 months.[3,4] Unlike cutaneous melanomas, mucosal melanomas are challenging to identify and diagnose.[1] Histopathology has confirmed that our case was a mucosal melanoma. Despite the typical absence of melanocytes in the colorectal mucosa, the reporting of mucosal melanoma cases has sparked curiosity about its pathogenesis. In the context of anorectal melanomas, the pathogenesis remains uncertain, with various theories proposed to explain their origin within the gastrointestinal tract. Some researchers hypothesize that anorectal malignant melanomas may arise from intestinal Schwann cells, while others propose an origin from neural crest cells that migrate during embryogenesis to the basal layer of the epidermis and hair follicles.[3] These differing perspectives underscore the complexity of melanoma

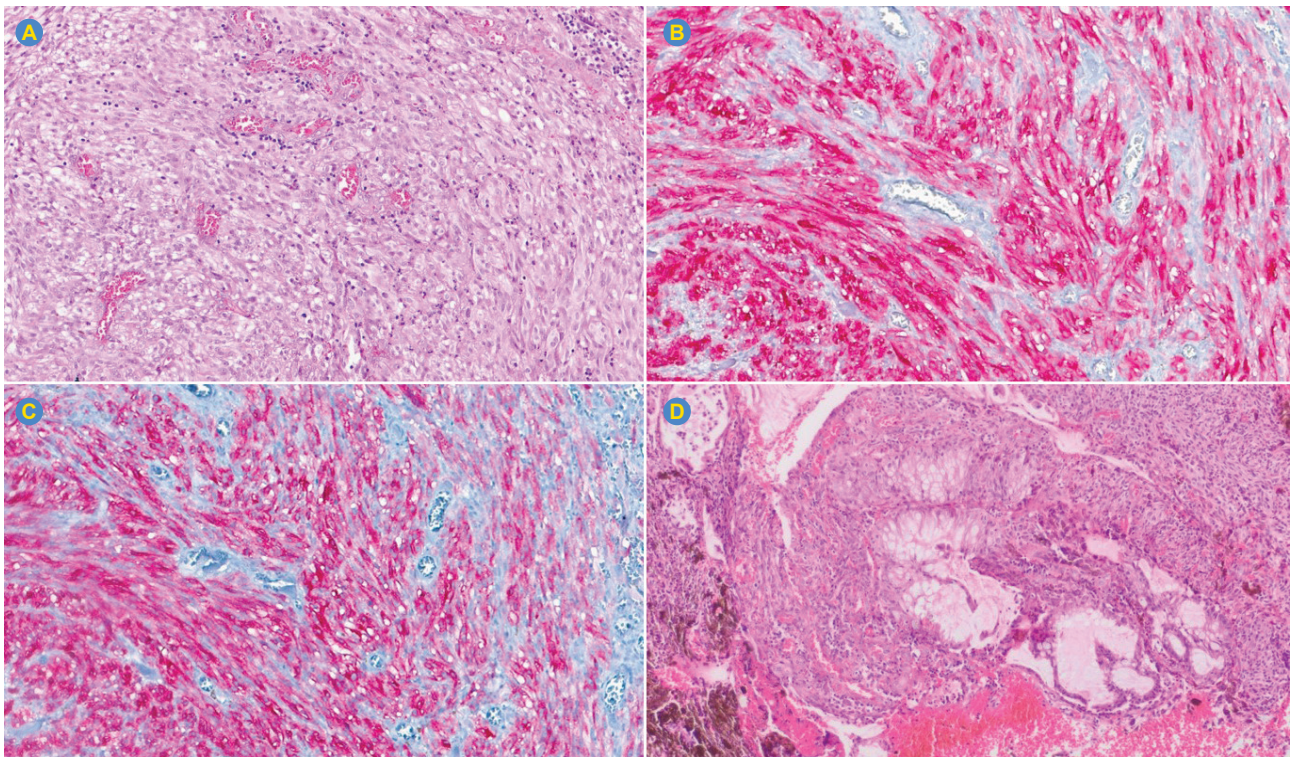

Figure 2 – Histopathological examination: tumoral infiltration composed of large hyperchromatic nuclei, distinct eosinophilic nucleoli, and pleomorphic cells (hematoxylin & eosin, x200 magnification) (A). Tumor cells diffusely positive for Melan A (immunohistochemical staining, AEC, x200 magnification) (B). Tumor cells diffusely positive for HMB 45 (immunohistochemical staining, AEC, x200 magnification) (C). Tumoral cells infiltrating the glands of the rectal mucosa (hematoxylin & eosin, x200 magnification) (D).

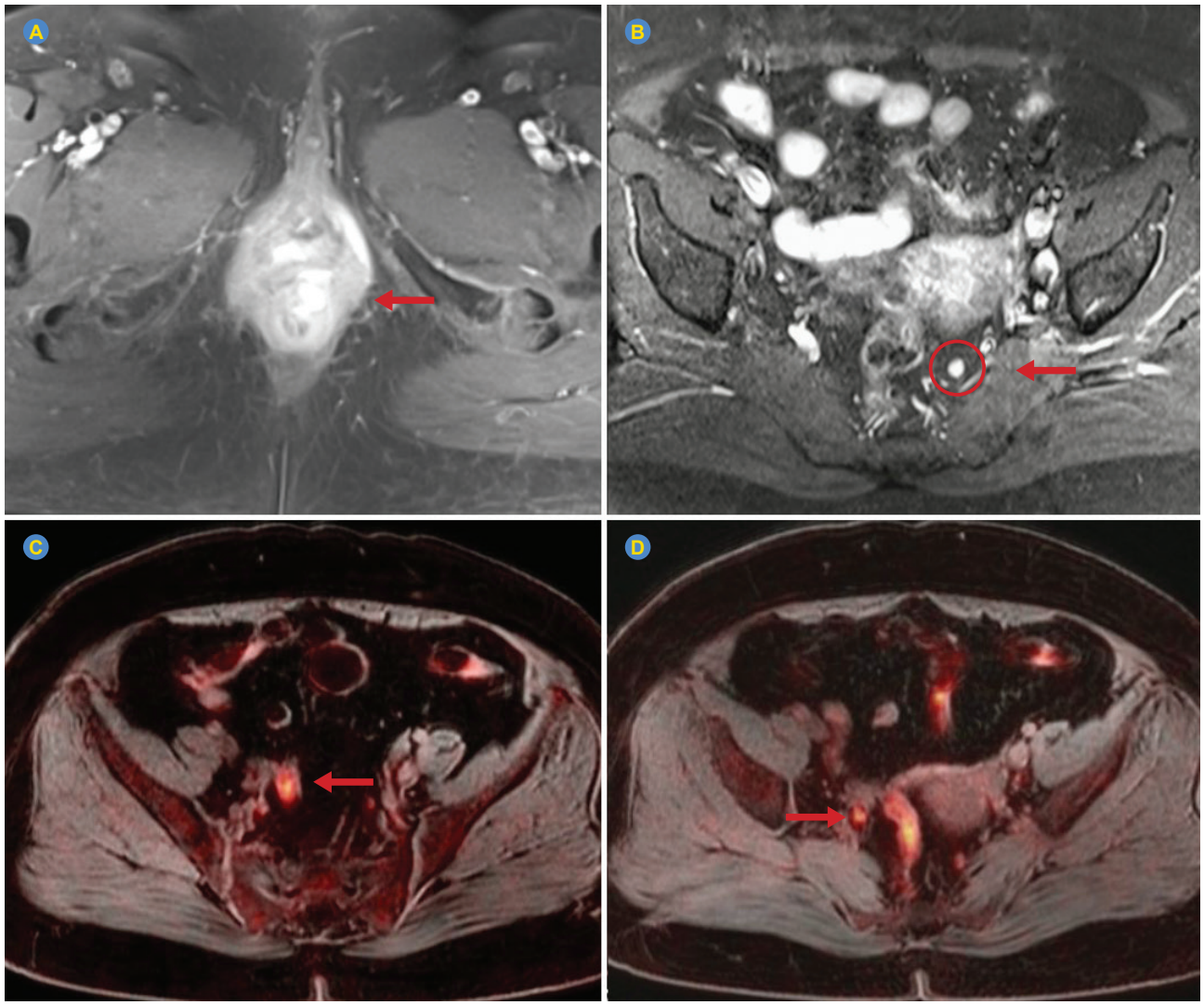

Figure 3 – Pelvic magnetic resonance imaging with T1 mapping: defect extending into the intersphincteric space (A). Perirectal lymphadenopathy (B). Positron emission tomography/magnetic resonance imaging: (C) and (D) pathological 18F-FDG uptake in the left presacral and left perirectal lymph nodes.

development and the need for further research to elucidate its underlying mechanisms.

Nonspecific symptoms and the absence of characteristic pigmentation in almost one-third of cases contribute to the suspected diagnosis of melanoma being overlooked.[1,2] Symptoms of anorectal malignant melanomas, located in approximately 65% of cases near the anal canal or anal verge, include bleeding, changes in skin color, changes in bowel habits, anorectal pain, or discomfort.[5] Hemorrhoids, polyps, adenocarcinomas, or rectal ulcers are erroneously diagnosed in nearly 80% of cases of primary anorectal malignant melanomas.[6] In our case, the most common symptom, rectal bleeding, accompanied by anal soiling and prolapse, was observed. However, since the initial examination was not performed at our hospital, the depigmented form cannot be confirmed definitively.

Anorectal melanomas often receive delayed diagnoses, and most patients have distant metastases not detected at the time of diagnosis.[2] Discovering extensive metastases in cases initially considered as easily treatable limited lesions is not uncommon. The diagnosis relies on clinical suspicion followed by biopsy.[1,3] The use of immunohistochemistry panels, including S-100 proteins, Melan-A, HMB-45, and tyrosinase, aids in the diagnosis.[7] Especially in amelanotic anorectal melanoma, careful pathological evaluation is necessary. In our case, HMB-45 and Melan-A were diffusely strongly positive.

Various imaging diagnostics are used in anorectal melanoma for the assessment of primary cancer, metastasis, and treatment responses. Endoscopic ultrasonography

(EUS) is valuable in evaluating lesion size, invasion depth, and extent, but in our case, MRI was preferred due to the presence of an open wound post-hemorrhoidectomy and the unavailability of EUS. Magnetic Resonance Imaging is highly effective in preoperative staging and detecting metastatic lesions.[3] In contrast, computed tomography (CT) excels in identifying hepatic and pulmonary metastases.[3] Due to its superior tissue characterization and soft-tissue resolution compared to CT scans, MRI stands out as the optimal modality for this condition. Positron emission tomography is recommended, particularly in the staging and assessment of treatment response in metastatic melanoma.[3] Increased FDG activity in malignant cells, due to their higher metabolic rates, allows PET to assess metastatic sites, aiding in staging and treatment decisions. In our case, the increased involvement observed in the anal canal and perirectal lymph nodes played a crucial role in determining the surgical strategy.

There is no standard approach for treating anorectal melanoma. Surgery, radiotherapy, and combinations of chemo-immunotherapy are used. Although surgery is the mainstay treatment, there is no consensus on the extent of resection and lymphadenectomy.[2] The rarity of this condition makes conducting randomized controlled trials challenging. Therefore, evidence from limited case presentations or case series is available.[4] Surgical treatments include APR and wide local excision (WLE). While WLE stands out with low morbidity and preservation of sphincter function, local recurrences are common. Despite the aggressiveness of APR, the likelihood of local recurrence is lower.[3,4,8] The perspective of advocating WLE as the primary treatment assumes that documented local recurrences do not impact survival.[8] Additionally, it has been reported that WLE provides equivalent oncological outcomes to APR, and APR is recommended, especially for locally advanced, large tumors, and WLE recurrences.[9] Based on clinical and radiological findings, and considering the patient's treatment preferences, we performed APR, traditionally deemed the optimal choice for regional disease management. Undoubtedly, in this approach, radiological lymph node positivity and the patient's decision were the most influential factors. Ultimately, although the postoperative pathological examination revealed no residual tumor, the presence of metastatic lymph nodes was identified.

In conclusion, when evaluating patients with symptoms related to the anorectal region, consideration should be given to rare and unusual conditions, such as malignant melanoma, in addition to typical benign lesions or adenocarcinoma when making a differential diagnosis.

AUTHOR CONTRIBUTIONS

RK: Conception and design of the work, data acquisition, analysis, and interpretation, drafting of the work, critical review, approval of the final version of the manuscript.

OA, MT: Data acquisition, analysis, and interpretation, approval of the final version of the manuscript.

All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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