

Case Report

Malignant Melanoma of Anorectal: A Report of Two Cases

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ABSTRACT

Introduction and importance: *Occurring in only 1% of cases of anorectal malignancy, malignant melanoma generally appears in the fifth and sixth decades of life, with complaints of anorectal bleeding or pain. The prognosis is generally poor. Although anorectal melanoma currently lacks a recommended treatment, surgery alone remains the primary modality of treatment, the role of adjuvant therapy is generally minimal, and the survival rate improves with early diagnosis.*

Presentation of case: *Here, we report two cases of a rare malignant melanoma in the rectum treated with abdominoperineal resection and local excision. The first case is a 60-year-old man with a history of defecating bloody stool and the appearance of a lump on the anal opening. The abdominoperineal resection was performed followed by adjuvant chemoradiotherapy. The second case is a 51-year-old woman with a similar complaint and clinical finding to the first case. The second case was treated with a tumor excision procedure followed by adjuvant chemoradiotherapy. However, the lump was recurrence after 7 months of prior tumor excision.*

Discussion: *Surgical resection of anorectal melanoma is debatable. While surgical resection is still the most common treatment option, there is no agreement on*

whether abdominoperineal resection or local excision is the best option. **Conclusions:** The abdominoperineal resection may offer a lower recurrence probability than that of tumor excision alone. However, further study is required to provide evidence-based surgical treatment.

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1. INTRODUCTION

Anorectal melanoma is a malignant tumor arising from melanocyte cells that develops in the anal canal.^{1,2} Metastasis from malignant melanoma (MM) is known to spread by local extension, by the lymphatics, or by the bloodstream.³ Most commonly appearing on the skin, followed by the meninges, eyes, mucosae, anorectal region, and the bone⁴, melanoma occurs with the same incidence in men as in women, especially ones who have experienced frequent exposure to sunlight.² Among men, melanoma most often appears on the upper back, whereas women most often experience it on the lower legs.⁵ Regardless of gender, adults and especially older adults contribute most to melanoma's incidence, with a peak among people in the sixth decade of life.⁶

Extremely rare, anorectal melanoma is responsible for only 1% of cases of anorectal malignancy, which is almost always caused by adenocarcinoma and various squamous cell cancers. Predominantly occurring among women, anorectal melanoma is most common in the fifth and sixth decades of life, and patients with the disease usually present with rectal bleeding and an altered defecation pattern. The prognosis for anorectal melanoma is generally poor, with a median survival of 24 months and a 5-year survival rate of 10%.² Most people with melanoma of the rectum die from metastases. Although no consensus currently exists regarding the most appropriate surgical approach for anorectal melanoma, possible procedures range from local excision to abdominoperineal resection (APR), either with or without adjuvant radiotherapy.^{2,7} Here, we report two cases of a rare malignant melanoma in the rectum treated in our institution.

2. CASE PRESENTATION

Case 1

A 60-year-old man with a 6-month history of bloody stool was admitted to the hospital with complaints of painful lumps in the anal opening 2 days prior to admission. On physical examination, the patient looked pale, no abnormality was detected by abdominal examination, and indicated no pain but also presented a brittle mass in the anus measuring 7 × 7 cm, with the consistency of dense rubber and that bled easily (Figure 1). Digital rectal examination suggested that the mass had originated on the anal verge.

Beyond that, laboratory tests revealed a hemoglobin (Hb) count of 7.4 g/dL and a Carcinoembryonic antigen (CEA) count of 8.5 ng/ml, and the results of a biopsy indicated malignant melanoma. Next, a computerized tomography (CT) scan showed a mass in the rectum but no lymph nodes (Figure 2) or metastases in the liver, and the chest X-ray did not reveal any metastatic nodules in the lungs. Considering the symptoms, physical examination, radiological examination, and histopathology results,

we classified the first patient as cT3N0M0 (stage IIB) according to the American Joint Committee on Cancer (AJCC) 8th edition (AJCC 8).



Fig. 1 Mass of fragile anorectal tumor that bled easily

The patient was fully informed about the treatment modality and choice. The abdominoperineal resection (APR) was explained as a procedure that completely removes the distal colon, rectum, and anal sphincter complex using both anterior abdominal and perineal incisions, resulting in a permanent colostomy. The tumor excision procedure was also explained as an alternative surgery. We had also informed the complications of tumor excision procedure which were the higher risk of bleeding, infection, chronic wound, recurrence probability, and fecal incontinence. We are concerned about the quality of life and the autonomy of the patient. The patient understood completely the treatment and agreed to APR. In consideration of those results, the patient underwent APR with a good outcome and was discharged on the eighth day after surgery. Once the pathology result of malignant melanoma was confirmed, the patient was scheduled for adjuvant chemoradiotherapy (Figure 3A dan 3B).

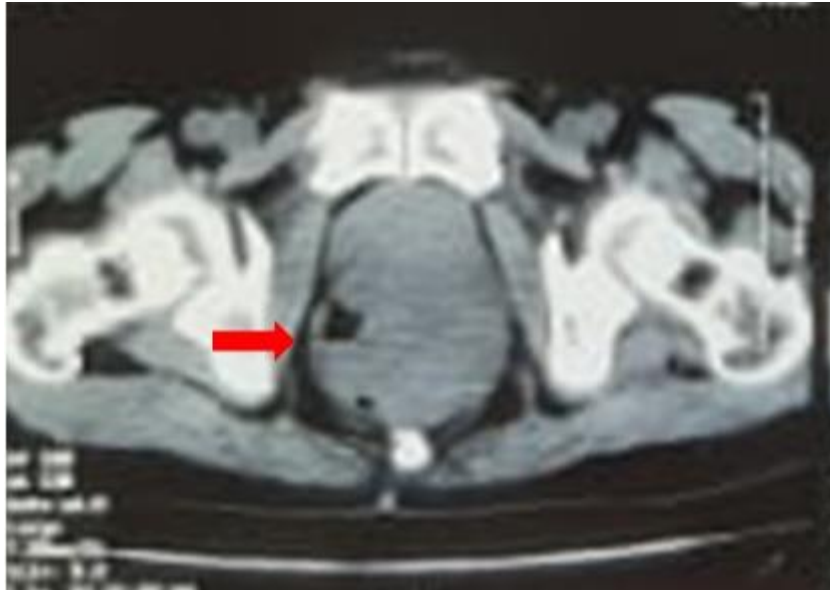


Fig.2. A CT scan of the abdomen and mass in the rectum (arrow).

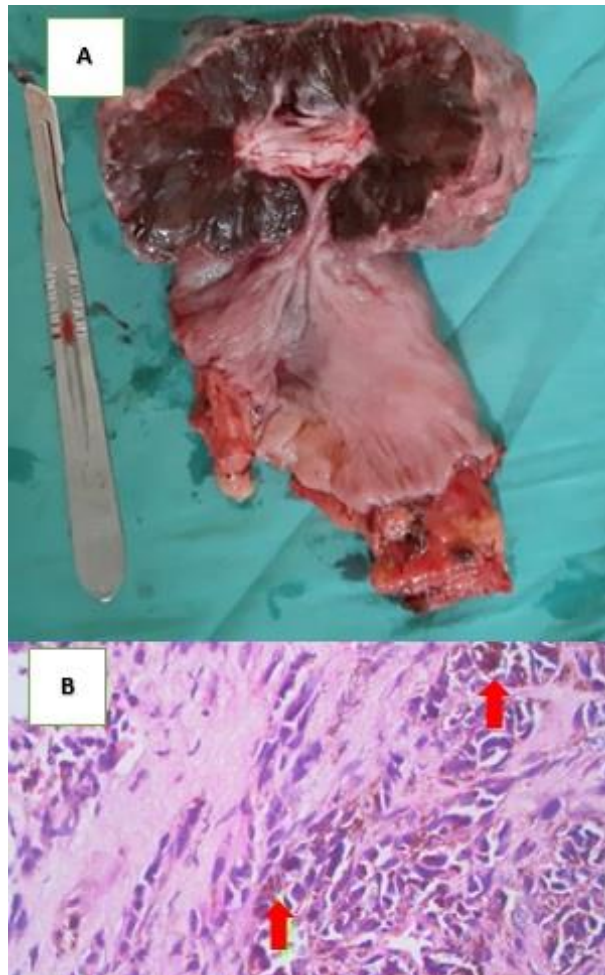


Fig. 3. a) Tumor mass after abdominoperineal resection; b) Histopathological results of 40x enlargement showing atypical cells with spindle nuclei contained melanin.

Case 2

A 51-year-old woman with a 1-year history of anorectal bleeding presented at the hospital with complaints of lumps in the anus for the past month. A year prior, the patient had received an endoscopy that revealed a polyp mass in the rectum indicating pathology of chronic inflammation (Figure 4). On physical examination, the patient looked pale, with no distention of the patient's abdomen, but also an anal mass measuring 10 × 15 cm that was fragile and bled easily. Laboratory tests revealed a Hb count of 6.6 g/dL, while the chest X-ray showed no signs of pulmonary metastatic nodules. We classified the second patient as cT3N0M0 (stage IIB) according to AJCC 8. The patient was well informed about the surgical treatment modality (APR and tumor excision) and the complication of the surgery. The patient chose the tumor excision procedure.



Fig. 4. Anorectal mass that is fragile and bleeds easily.

The tumor was excised, and its examination confirmed the pathology of malignant melanoma (Figure 5). The patient was discharged 6 days postoperative in good condition. The patient was completely informed about the adjuvant chemoradiotherapy, including the benefit and the complication. The chemoradiotherapy session had been scheduled from the outpatient clinic. However, the patient refused to have chemoradiotherapy immediately with the consideration of waiting for the surgical wound to heal. The patient went back to her hometown which was far from our institution.

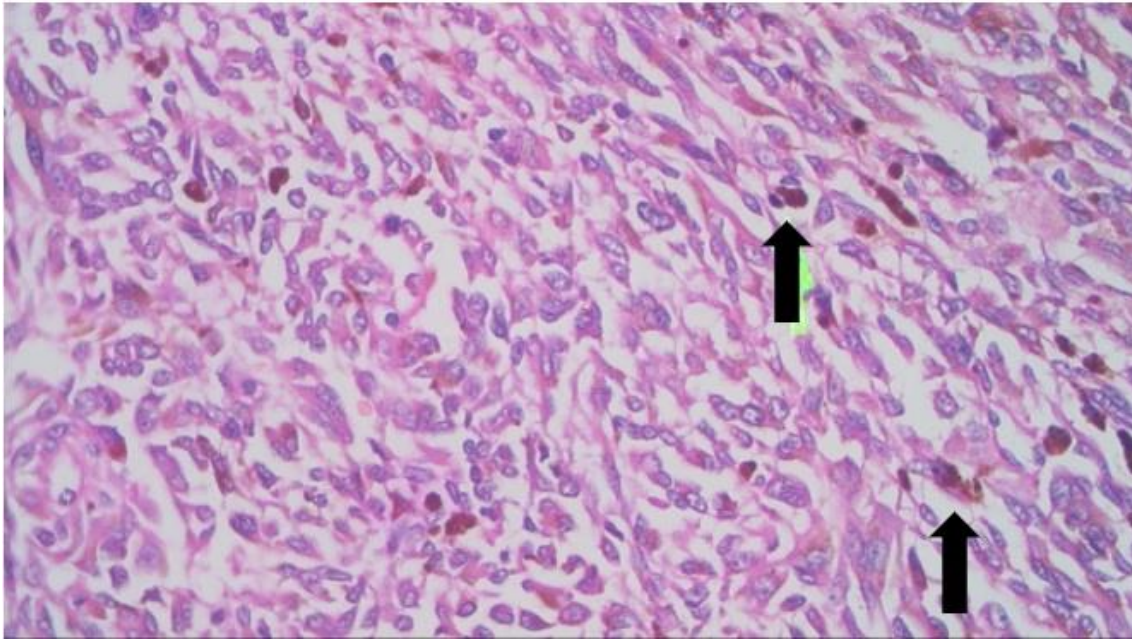


Fig.5. Histopathological results of 40x enlargement show atypical cells with spindle nuclei containing melanin.

Seven months later, the patient returned with new complaints of anal bleeding without any prior adjuvant chemoradiotherapy, which general conditions Karnofsky grade 60%. A physical examination of the patient revealed an anemic, flat, but painless abdomen, after which a digital examination of the rectum revealed a mass measuring 2 x 5 cm at the base of the dentate line that bled easily. A CT scan of the abdomen showed a rectal mass with multiple metastatic nodules on the liver, and a chest x-ray showed metastases in the lungs. A re-excision of the tumor was performed to stop the bleeding, and the patient was discharged on the fourth day after surgery.

3. DISCUSSION

First reported by Moore in 1957, anorectal melanoma is a highly rare disease. Melanoma of the rectum, which accounts for just 1% of anorectal malignancies and occurs predominantly in women in their fifth and sixth decades of life, is difficult to detect and is sometimes misdiagnosed as rectal polyps, hemorrhoids, or ulcer lesions that co-occur with rectum prolapse.⁸

Anorectal melanoma appears on the dentate line, where it develops from melanocytes in the dentate area. From a macroscopic standpoint, the tumors are polyp-shaped, and if microscopically pigmented with immunostaining features for melanosomal proteins, the cells appear to be arranged in nests. The histological markers S-100, HMB-45, and vimentin can help to clarify the diagnosis. If the biopsy shows a suspicious specimen for a sarcoma (e.g., leiomyosarcoma), then S-100 staining should be performed, and if the result is positive, then the tumor is most likely melanoma.⁹ Tumor histology includes epithelial cells or spindles; however, as in our second case, histology results can be a mixed tumor, round oval to the spindle. Tumors with pure epithelioid histology are less likely to recur.²

The rarity of malignant melanoma of the rectum stems from the observation of tubulovillous polyps in the tumor pedicles. Melanoma occurring at the anal verge can turn into a polyp and become an invasive melanoma in the rectal mucosa, after which infiltration into the dysplastic polyp occurs. For that reason, such melanoma is also sometimes called a “collision tumor”.⁹

Wide local excision (WLE), abdominoperineal resection (APR), and endoscopic mucosal resection (EMR) are some of the surgical options. In rare circumstances, EMR can resect the melanoma and provide long-term survival. WLE has low morbidity and does not impair local function while preserving the anal sphincter. APR is frequently linked to a high risk of morbidity and functional impairment. When the tumor margins were macroscopically and microscopically negative, the operation was classed as R0, R1 when the margins were positive on microscopy, and R2 when the resection was macroscopically incomplete.¹

Surgical resection of anorectal melanoma is debatable. While surgical resection is still the most common treatment option, there is no agreement on whether APR or local excision is the best option. Because it can restrict lymphatic dissemination and provide a bigger negative margin for local control, APR is considered the standard procedure for treating anorectal melanoma.⁹

Maliha Khan et al reported a case of a 71-year-old female with anorectal melanoma. Their patient was treated with APR prior to chemotherapy. However, the long-term outcome was unknown.¹⁰ In 2019, Binh Van Pham et al reported two cases of anorectal melanoma. One of their cases was treated with APR and another with local excision of the tumor followed by ultra-low anterior resection (ULAR). The patient who underwent the APR procedure had no complications from surgery and no sign of metastases or recurrence 6 months after surgery, whereas the patient who received the ULAR experienced low anterior resection syndrome and evidence of recurrence was found 16 months after the surgery.¹¹

Despite the lack of consensus on whether rectal melanoma should be treated with APR or wide local excision,¹² several recent studies have suggested that, if possible, sphincter-sparing local excision and adjuvant radiation can effectively control loco-regional disease while preventing functional morbidity due to APR. At the same time, mesorectal lymph nodes play a role in the appearance of inguinal lymph nodes, which differ in incidence from squamous cell carcinoma in the anus. Thus, if an APR is performed, then mesorectal lymph node resection can contribute to better staging, whereas prophylactic inguinal lymph node resection will have no benefit. As for the outcomes, patients without lymph node metastases had a 5-year survival rate of 20% versus 0% among patients with lymph node metastases. Meanwhile, the survival rate of patients with recurrent or metastatic rectal melanoma is less than 10 months.⁹ Poor prognosis in tumors is influenced by tumor thickness, tumor necrosis, and perineural invasion, and amelanotic lesions typically receive worse prognoses.¹³

4. CONCLUSION

Malignant melanoma of the rectum, a rare anorectal malignancy, is a highly aggressive disease that is difficult to diagnose. Although anorectal melanoma currently lacks a recommended treatment, surgery alone remains the primary modality of

treatment, the role of adjuvant therapy is generally minimal, and the survival rate improves with early diagnosis.

INFORMED CONSENT

Written informed consent was obtained from the patients for participation in our study.

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Conflict of Interest Statement:

The author declares that the case report was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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