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CASE REPORT

Malignant Melanoma Anal Canal— A Rare Location

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Abstract

Anorectal malignant melanoma (AMM) is rare, presents a diagnostic challenge often confused with rectal carcinoma, which constitutes the majority of rectal tumors. Its aggressive nature leads to a poor prognosis with fatal outcomes. Early diagnosis is crucial due to its nonspecific symptoms. Combination of complete surgical resection with adjuvant chemotherapy and immunotherapy may be of benefit. Here, we present one such case managed successfully by abdominoperineal resection (APR).

Key Words

AMM, APR, Surgical Resection

Introduction

Anorectal malignant melanoma (AMM), a rare and aggressive melanoma subtype, was first reported by Moore et al. in 1857.[1] Although it is the third most common site after cutaneous and ocular melanomas, anorectal malignant melanoma accounts for less than 2% of all melanoma cases and makes up 24% of mucosal melanomas.[2,3] Furthermore, it represents a minority of anal tumors, accounting for less than 2%.[4] Typically diagnosed in advanced stages, AMM presents notable clinical complexities.

Due to the limited effectiveness of chemotherapy and radiotherapy, surgery is the primary treatment for AMM. However, there is ongoing debate about the best surgical strategy, particularly concerning the extent of resection—wide local excision versus abdominoperineal resection.

AMM has a poor prognosis, marked by high recurrence and mortality rates. The 5-year survival rate is below 20%, and median survival is under 2 years.[1,2,4,5]

Here, we present a one such case of amalignant melanoma of anal canal and its treatment.

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Case Report

A 67 year gentleman presented with a history of bleeding per rectum on and off for 2 years associated with painful defecation, tenesmus and significant weight loss with no obstructive features. On per rectal examination a polypoidal mass was felt over the anterior wall of anal canal, from 7-1 o'clock position just above the anal verge.

Biopsy of the lesion: malignant melanoma, positive for HMB45, Melan A, S-100 while negative for p40.All blood investigations including CEA were in normal range.

CECT abdomen and pelvis: Mild diffuse edematous neoplastic wall thickening involving terminal anal canal, extending into external anal opening. (Figure 1)

MR Pelvis-T1 and T2 weighted images showing well-defined, lobulated, mass lesion occupying upper half of the anal canal. Upper margin of the lesionreaching upto anorectal junction, encircling 3/4th of the lumen, extending from 2o'clock to 11 o'clock positions causing significant luminal compromise, infiltrating and reaching upto serosal surface, predominantly along the right side. The

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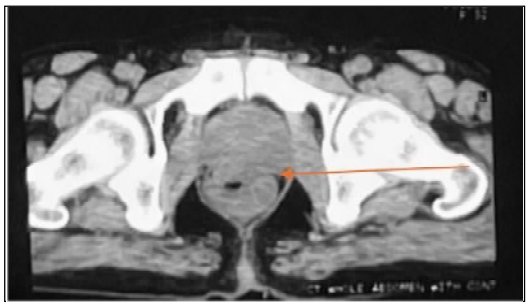


Figure 1: CECT abdomen and pelvis showing a growth and thickening of the anal canal with luminal compromise.

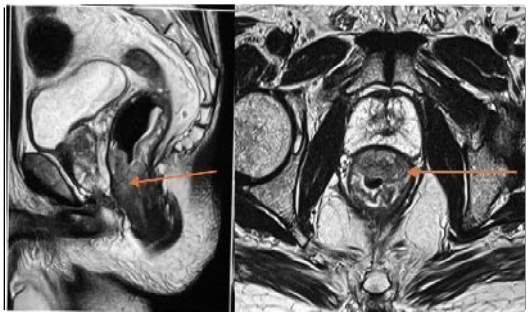
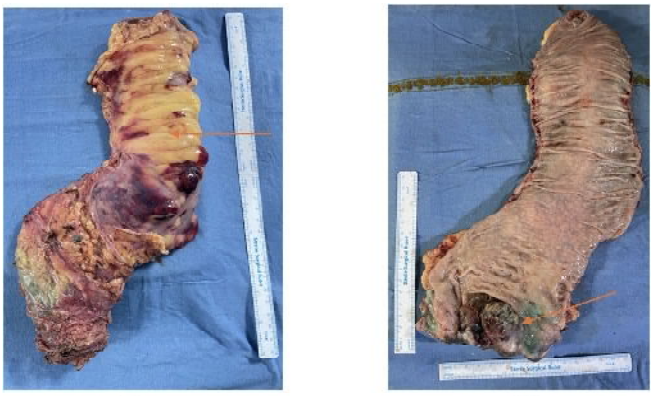


Figure 2: MRI pelvis (a) Sagittal, (b) Axial sections showing anal growth occluding 3/4th of the lumen.



(a) (b)

Figure 3: Specimen images (a) Total mesorectal excision showing intact mesorectum with no irregularities and coning, (b) Cut section showing a polypoidal ulcerated growth at the anal verge.

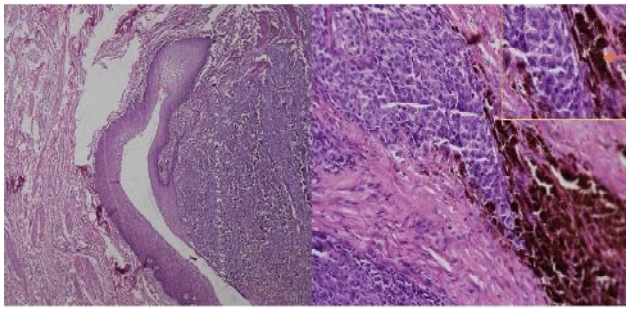


Figure 4: Histopathology (a) Tumor cells arranged in nests. A few of these nests prominently display melanin pigmentation. (20X) (b) A microscopic view of the anal canal reveals malignant melanoma originating from the squamous epithelium, with tumor cells exhibiting enlarged nuclei, irregular chromatin, and prominent eosinophilic nucleoli (10X, 40X).

surrounding mesorectal fat planes preserved. (Figure 2a and 2b)

PET Scan- FDG avid irregular circumferential wall thickening is noted involving anal canal and adjacent rectum (max thickness~2.5 cm, length~3.8 cm). Fat planes with prostate is lost anteriorly. FDG avid tiny mesorectal lymph node is noted on right side.

USG Inguinal region: Few lymph nodes seen in right and left inguinal region with maintained fatty hilum measuring 18 x 5 mm on right and one of them on the left side shows loss of fatty hilum and measures 5 x 3 mm. Rest show maintained fatty hilum.

Patient underwent laparoscopic APR.

One day prior to the planned procedure and before starting the procedure, Technetium colloid dye 10 ml injection in the peri-tumoral region was done. Just before the surgery peri tumoral injection of the methylene blue dye was also done. No bilateral inguinal lymph nodal uptake of technetium sulphur colloid - confirmed using gamma probe. There was no evidence of metastasis. (Figure 3a and 3b)

Post -operative recovery was uneventful. Six months post surgery, patient does not have any recurrence.

Histopathology: Malignant melanoma arising from the anal skin, infiltrating into the muscularis propria with no lymphovascular and perineural invasion seen. Resected margin free of tumor. Thirteen mesorectal lymph nodeswere reactive. (0/13). **Stage:** pT3 pN0. (Figure 4a and 4b)

Discussion

AMM is a rare and aggressive melanoma subtype with a poor prognosis, high recurrence and mortality rates. First described by Moore *et al*. in 1857, representing less than 2% of all melanomas and 24% of mucosal melanomas.[1,2,3] Typically diagnosed in the sixth to seventh decade of life, often presents with non-specific symptoms, leading to late-stage detection with sizable masses and widespread disease.[4] Metastasis commonly

occurs in the inguinal, mesenteric, hypogastric, and para-aortic lymph nodes, with distant spread often involving the liver, lungs, brain, and bones.[4]

The 5-year survival rate is less than 20%, with a median survival of fewer than 2 years.[5]

Diagnostic assessment comprises of colonoscopy, endoscopic ultrasound and imaging such as thoracic, abdominal and pelvic CT or PET-CT scans. Histological markers like S-100, HMB-45 and Vimentin assist in diagnosis.[4,6]

Surgical intervention is the preferred treatment. Prophylactic inguinal lymph node dissection does not improve survival rates and only increases surgical morbidity, even with clinically positive nodes.[1,4]

There is no established standard systemic therapy for AMM. However, combining complete surgical resection with adjuvant chemotherapy like dacarbazine, vinblastine, cisplatin, immunotherapy such as pembrolizumabmay confer better survival compared to surgery alone.[4,7] Immunotherapies are increasingly being explored for treating AMM, although research indicates that MMs generally respond less to immunotherapy compared to chemotherapy. No specific follow up guidelines exist for AMM. Patient can be kept on 6 monthly surveillance.

Conclusion

AMM is a rare, but an aggressive tumor. The clinical, biological, and molecular variances distinguish mucosal melanoma significantly from its cutaneous counterpart. A good complete surgical resection is the mainstay of treatment.

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