# Case Report

DOI: https://dx.doi.org/10.18203/2349-2902.isj20211838

# Two case reports of malignant melanoma rectum

## Jeena K. Sathyan\*, Manmohan Kamat, Varsha Sharma, Shravani Shetye, Seema Barman

Department of General surgery, Nanavati superspeciality hospital, Mumbai, Maharashtra, India

Received: 12 January 2021 Revised: 23 February 2021 Accepted: 18 March 2021

## \*Correspondence: Dr. Jeena K. Sathyan,

E-mail: jeenaks014@gmail.com

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

#### **ABSTRACT**

Malignant melanoma of the rectum is an extremely rare disease. It typically presents in the fifth or sixth decade predominantly in female sex. The first symptoms are nonspecific such as bleeding, anal mass or pain. A timely diagnosis of melanoma is made even more difficult due to lack of obvious pigmentation and histologically amelanotic. Anorectal malignant melanoma spread along submucosal planes and are often beyond complete resection at the time of diagnosis. Prognosis is very poor. We present a rare case of malignant melanoma of rectum in a 21-year-old male, who was diagnosed at advanced stage, and a case of malignant melanoma in a 50-year-old male, who underwent abdominoperineal resection with permanent colostomy. Anorectal malignant melanoma is difficult to diagnose and prognosis is poor.

**Keywords:** Malignant anorectal melanoma, Rare case, Abdomino-perineal resection, Adjuvant chemo-radiotherapy, Colostomy

## INTRODUCTION

Malignant melanoma of rectum is extremely rare, and very aggressive disease. It constitutes to about 0.5-4% of all anorectal malignancies and less than 1% of all melanomas.<sup>1,3</sup> More common in women, typically presents in the fifth or sixth decade of life.<sup>4</sup> Strong association with Caucasian race. Prognosis is very poor with median survival of 24 months and 5-year survival of 10-15%.6 A timely diagnosis of anal melanoma is made even more difficult by the fact that up to 80% of lesions lack obvious pigmentation and up to 20% of tumors are even histologically amelanotic.7 Due to rarity of this malignancy there is no consensus on which surgical approach is favorable, also about the adjuvant therapy available. The surgical procedure of choice ranges from an Abdominoperineal resection (APR) to Wide local excision (WLE) with or without adjuvant radiotherapy.

Here are two case reports of malignant melanoma rectum who was managed in our institute.

### **CASE REPORT**

## Case 1

We reported a case of 21-year-old male patient, presented with c/o passing clots/bleeding per rectal since 1year (04 June 2019). Fleshy growth on rectal examination. Biopsy was taken from the same and was referred to surgical oncology department with the following report. Other systemic examination was essentially normal.

On 10 June 2019: Histopathology report suggesting: Amelanotic Malignant Melanoma, Ki 67 index high, HMB45- Positive, Vimentin- strongly and diffusely positive, Melanin A- strongly and diffusely positive. Patient underwent MRI pelvis on 13 June 2019- which reported, a large lobulated heterogeneously enhancing circumferential soft tissue mass is seen in mid and lower rectum which involves pelvic side walls and internal

sphincter. Few prominent lymph nodes are also seen in mesorectal fat and in pre-sacral space. With a Stage cT4b-N2.

On 15 June 2019, patient underwent CT Abdomen and pelvis which reported a large lobulated heterogenous enhancing soft tissue mass seen in rectum, extends upto lateral pelvic wall measuring 7.9×7.1 cm (axial plane) and 9.2 cm (superoinferior dimension). Few non-necrotic lymph nodes seen in mesorectal fat and in pre-sacral space 2×1.5 cm appears to be metastatic. No liver metastasis or upper abdominal lymphadenopathy.

Patient was deferred surgery by surgical oncology team due the advanced stage of the disease. And was referred to medical oncologist.

Diagnosis of advanced amelanotic malignant melanoma was made and further planned for PET CT, immunotherapy/chemotherapy. Patient came to Nanavati hospital (24 June 2019) with c/o pain in abdomen, burning micturition and poor stream of urine. With c/o partial obstruction with constipation for 1 month. Flatus tube was attempted but failed, reference to surgery department was given for emergency exploratory laparotomy. Patient underwent diagnostic laparoscopy with diversion colostomy for distal obstruction. Days. Patient was discharged on 12 July 2019. Patient was admitted again twice during 29 June to 02 August 2019 and 19 august to 24 august 2019 intraoperative findings were of dialated bowel loops++, sigmoid colon identified looped for a diversion colostomy. Patient tolerated the procedure well and was shifted to ICU. Further stay in hospital was uneventful. Chemotherapy started with dacarbazine (500 mg), given for 5 for chemotherapy. Patient had recurrent blockage of urinary catheter and acute pain in abdomen. Patient was admitted for third cycle of chemo when he had an episode of drowsiness and was shifted to ICU, where he had one episode of seizure and not responsive. MRI brain was done and was so meningeal metastasis. Patient succumbed in the same admission.

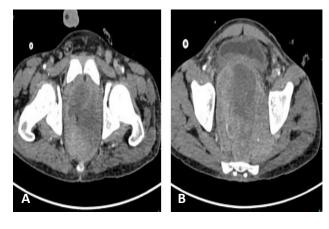


Figure 1: Contrast-enhanced axial CT scan images of pelvis shows heterogenous mass lesion in rectum with extension into mesorectal fat.

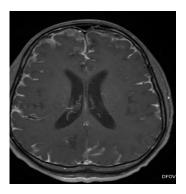


Figure 2: Leptomeningeal metastasis.

#### Case 2

We reported a case of 50-year-old male, came on 19 December 2019 with complaints of bleeding per rectum. Which was diagnosed as polyp underwent colonoscopy followed by haemorrhoidectomy on 21 November 2019, biopsy was s/o malignant tumor invading into muscle layer- malignant melanoma- poorly differentiated carcinoma.

PET-CT done on 10 December 2019 suggestive of sessile polypoidal lesion left lateral wall of lower rectum 1.7×1.9×1.5 cm lies 5 cm away from anal verge, transmural infiltration, perirectal stranding, increased left internal iliac LN- SUV max 7.64.

Past history of the patient was hypertension and type II Diabeties mellietus. Patient underwent abdominoperineal resection with permanent colostomy. Following which he was subjected to radiotherapy. Patient is on regular follow up, no evidence of recurrence.

# **DISCUSSION**

Anorectal malignant melanoma (ARM) neuroectodermal neoplasm originating from melanoblastic cells of mucosal surface. During fetal development, these cells migrate throughout the body, primarily the skin. However, melanocytes also reside in the eyes (retina and uveal tract) and mucosal surfaces.<sup>3</sup> Therefore, cutaneous melanomas are by far the most common form of the disease, comprising more than 90% of all melanomas. Of the remaining (<10%) forms of melanoma, ocular melanoma accounts for 5%, melanoma of unknown origin for 2%, and mucosal melanoma for 1%.8 'Primary anorectal melanoma' occurs around the dentate line. Macroscopically, the tumors are polypoidal and pigmented while microscopically, the cells are arranged in nests with characteristic immunostaining specific for melanosome protein. However, 30-70% of the lesions can be amelanotic. Malignant melanoma arising in anorectal lesions accounts for 0.4-1.6% of all malignant melanomas. ARM patients are more frequently female, and the median age at diagnosis is 60 years or higher. 10 Patients present with nonspecific complaints compared to other benign, or malignant disease of anorectum. The most frequent symptom is bleeding (54-78%), other symptoms include mass (12-16%), pain (14-27%), obstipation (6%), diarrhea (4%), and pathologic diagnosis after a hemorrhoidectomy (8-16%). Due to delayed diagnosis and the aggressive nature, 37% of the patients already have distant or regional metastasis at the time of diagnosis. The histologic markers of S-100, HMB-45 and vimentin are frequently identified and help clarify the diagnosis. 45,9

Lymphatic spread is common and tends to involve mesenteric and inguinal lymph nodes. The major sites of distant metastasis are lung, liver, and bone. Brain is the most common metastasis site, followed by liver and lung. The incidence rates for locoregional lymph node metastases on initial presentation are almost 60%. At the time of diagnosis, distant metastases are identified in 26-38% of patients. Anal melanoma is staged on a clinical basis, focusing on loco-regional and distant spread. Stage I is local disease only, Stage II is a local disease with increased thickness and ulcerations, Stage III is local disease with involvement of regional lymph nodes, and Stage IV shows distant metastatic disease. Stage IV shows distant metastatic disease.

A sigmoido-colonoscopy essential for evaluation of the cause of symptoms and obtaining a tissue biopsy from a suspicious lesion. Endoscopic endorectal ultrasound can be considered to evaluate tumor thickness and surrounding nodal status.<sup>5</sup>

Computed tomography (CT) scan of the abdomen and pelvis is valuable tool to assess regional disease, and is frequently used to see lymphadenopathy or metastasis.<sup>3,5,6</sup>

Contrast-enhanced CT scan and MRI allow characterization and assessment of the extent of the tumor. On CT scans, primary rectal malignant melanomas appear as bulky intraluminal fungating masses in the distal rectum, focally expanding and obscuring the lumen without causing obstruction, with perirectal infiltration and frequently enlarged lymph nodes. <sup>12</sup> Rectal carcinoma or other rectal masses present as significant obstruction.

MRI shows the melanotic component as high signal intensity on T1-weighted imaging and mixed signal intensity on T2-weighted imaging. Extraluminal extent of the lesion is better demonstrated by MRI imaging. Other rectal mass lesions show hypointense signal on T1-weighted imaging.

Treatment option has not been standardized due to low incidence and lack of evidence. Surgical excision is considered a primary treatment option. APR is considered as standard surgery, preferred because it can control lymphatic spread (mainly to mesenteric lymph nodes) and larger negative margin for local control. WLE (wide local excision) is less morbid, without stoma creation. Laparoscopic APR has the benefit of lesser morbidity.

Chemotherapy: Dacarbazine, Bacile Calmette Gurin (BCG), levamisole, cisplatin, vinblastine, intereukin-2 and interferon, have been investigated.

Radiotherapy: Adjuvant radiation therapy after surgical excision has also been attempted.

Radiotherapy to extended field followed by sphincter saving wide excision reduced locoregional recurrence rate to 17% from 50% compared to WLE alone.

#### **CONCLUSION**

From these two cases reports it's noted that young males have a poor prognosis due to delay in detection and early metastasis. Whereas the other patient is on regular follow up with no evidence of recurrence or distant metasis, However, long term surveillance is mandatory. More data is required and a treatment protocol is to be devised according to the presentation.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

## **REFERENCES**

- 1. Roviello F, Cioppa T, Marrelli D, Nastri G, De Stefano A, Hako L, et al. Primary ano-rectal melanoma: considerations on a clinical case and review of the literature. Chir Ital. 2003:575-80.
- Solaz Moreno E, Vallalta Morales M, Silla Búrdalo G, Cervera Miguel JI, Díaz Beveridge R, Rayón Martín JM. Primary melanoma of the rectum: an infrequent neoplasia with an atypical presentation. Clin Transl Oncol. 2005;7(4):171-3.
- 3. Singer M, Mutch MG. Anal melanoma. Clin Colon Rectal Surg. 2006;19:78-87.
- Schaik VPM, Ernst MF, Meijer HA, Bosscha K. Melanoma of the rectum: a rare entity. World J Gastroenterol. 2008;14(10):1633-5.
- 5. Stefanou A, Nalamati SP. Anorectal melanoma. Clin Colon Rectal Surg. 2011;24(3):171-6.
- Liptrot S, Semeraro D, Ferguson A, Hurst N. Malignant melanoma of the rectum: a case report. J Med Case Rep. 2009;3:9318.
- 7. Morson BC, Volkstädt H. Malignant melanoma of the anal canal. J Clin Pathol. 1963 Mar;16(2):126-32.
- 8. Row D, Weiser MR. Anorectal melanoma. Clin Colon Rectal Surg. 2009;22(2):120-6.
- 9. Reid A, Dettrick A, Oakenful C, Lambrianides A. Primary rectal melanoma. J Surg Case Rep. 2011;2011(11):2.
- 10. Ballo MT, Gershenwald JE, Zagars GK, Lee JE, Mansfield PF, Strom EA, et al. Sphincter-sparing local excision and adjuvant radiation for anal-rectal melanoma. J Clin Oncol. 2002;20(23):4555-8.

- 11. Carcoforo P, Raiji MT, Palini GM, Pedriali M, Maestroni U, Soliani G, et al. Primary anorectal melanoma: an update. J Cancer. 2012;3:449-53.
- 12. Kim KW, Ha HK, Kim AY, Kim TK, Kim JS, Yu CS, et al. Primary malignant melanoma of the rectum: CT findings in eight patients. Radiology. 2004;232(1):181-6.
- 13. Matsuoka H, Nakamura A, Iwamoto K, Sugiyama M, Hachiya J, Atomi Y, Masaki T. Anorectal malignant melanoma: preoperative usefulness of

magnetic resonance imaging. J Gastroenterol. 2005;40(8):836-42.

Cite this article as: Sathyan JK, Kamat M, Sharma V, Shetye S, Barman S. Two case reports of malignant melanoma rectum. Int Surg J 2021;8:1603-6.