



# International Journal of Surgery Science

E-ISSN: 2616-3470

P-ISSN: 2616-3462

© Surgery Science

[www.surgeryscience.com](http://www.surgeryscience.com)

2022; 6(3): 04-08

Received: 09-04-2022

Accepted: 16-05-2022

## Rashpal Singh

Assistant Professor, Surgical  
Oncology, Cell in the Department  
of General Surgery, Indira Gandhi  
Medical College, Shimla, Himachal  
Pradesh, India

## Puneet Mahajan

Professor, Department of General  
Surgery, Indira Gandhi Medical  
College, Shimla, Himachal  
Pradesh, India

## Vivek Rajdev

Junior Resident, Department of  
General Surgery, Indira Gandhi  
Medical College, Shimla, Himachal  
Pradesh, India

## Jagwinder Singh

Junior Resident, Department of  
General Surgery, Indira Gandhi  
Medical College, Shimla, Himachal  
Pradesh, India

## Venkatesh Singhal

Junior Resident, Department of  
General Surgery, Indira Gandhi  
Medical College, Shimla, Himachal  
Pradesh, India

## Kartik Rana

Junior Resident, Department of  
General Surgery, Indira Gandhi  
Medical College, Shimla, Himachal  
Pradesh, India

## Corresponding Author:

### Rashpal Singh

Assistant Professor, Surgical  
Oncology, Cell in the Department  
of General Surgery, Indira Gandhi  
Medical College, Shimla, Himachal  
Pradesh, India

## Anal canal melanoma-a rare entity: A case report and review of literature

**Rashpal Singh, Puneet Mahajan, Vivek Rajdev, Jagwinder Singh,  
Venkatesh Singhal and Kartik Rana**

DOI: <https://doi.org/10.33545/surgery.2022.v6.i3a.907>

### Abstract

Anorectal mucosal melanoma is one of the rarest neoplasm and accounts for approximately 0.05 percent of all colorectal malignancies and 1 percent of all anal canal cancers. Patients typically present with bleeding, a mass, anorectal pain, or a change in bowel habits. Management includes surgical resection depending upon the site along with adjuvant chemotherapy, radiotherapy and immunotherapy.

48 years old gentleman who presented with 6-month-old history of bleeding per rectum, pain and tenesmus. On digital rectal examination, a semi-circular polypoidal growth was present posteriorly 2 cm from anal verge with cranio-caudal extent of 5 cm. Biopsy was suggestive of malignant melanoma. MRI pelvis revealed asymmetric circumferential thickening of 30 mm for length of 3.8 cm located 3.4 cm from anal verge with enlarged mesorectal and extra-mesorectal lymph nodes. Patient was planned for abdominoperineal resection. Surgery and post-operative period were uneventful.

Final histopathology revealed polypoidal tan-brown, partly ulcerated growth of 4 x 2.5 x 2 cm on cut-section. These findings were consistent with malignant melanoma. All 15 lymph nodes dissected were free from tumor deposits.

**Keywords:** Melanoma, anal canal, case report

### Introduction

Anorectal mucosal melanoma is one of the rarest neoplasm and accounts for approximately 0.05 percent of all colorectal malignancies and 1 percent of all anal canal cancers<sup>[1]</sup> The site of origin is the rectum or anal canal in 42 and 33 percent of cases, respectively, while the primary site cannot be determined in the remainder. Although the risk factors for anorectal mucosal melanoma are not known, epidemiologic data suggest that there is an increased risk associated with human immunodeficiency virus infection<sup>[2, 3]</sup>.

The majority of cases arise from the mucocutaneous junction; however, they can also arise from the skin of the anal verge, the transitional epithelium of the anal canal, or the rectal mucosa. Identification of the primary site of disease is critical to separate anal cutaneous from anal mucosal melanomas, as both of them differ in terms of management<sup>[4]</sup>.

Patients typically present with bleeding, a mass, anorectal pain, or a change in bowel habits<sup>[5, 6]</sup>. Occasionally, melanoma is an incidental finding on pathologic evaluation of a haemorrhoidectomy or anal polyp specimen. Anorectal melanoma is pigmented in only one-third of cases<sup>[7]</sup>. Most patients present with lesions that are >2 mm thick<sup>[8]</sup>. Regional lymph node involvement is found in approximately 60 percent of patients at presentation, and distant metastases are present at diagnosis in approximately 30 percent of cases<sup>[9, 10]</sup>.

The initial evaluation of patients with anorectal melanoma should include a rectal examination with punch biopsy and immunohistochemistry, rectal ultrasound, and CT and/or PET imaging to assess for distant metastases. Management includes surgical resection, which can vary from wide local excision to abdominoperineal resection depending upon the site, size and extent along with adjuvant chemotherapy, radiotherapy and immunotherapy<sup>[11]</sup>.

Our article describes a case report of a patient of anal canal mucosal melanoma along with review of literature that adds to our knowledge of management of mucosal melanoma.

### Case Presentation

48 years old gentleman who presented with 6-month-old history of bleeding per rectum, pain

and tenesmus. On digital rectal examination, asemi-circular polypoidal growth was present posteriorly 2 cm from anal verge with craniocaudal extent of 5 cm. Biopsy was suggestive of malignant melanoma and immunohistochemistry was positive for Melan-A, S100, HMB-45 and SOX-10, confirming the diagnosis. MRI pelvis revealed asymmetric circumferential thickening of 30 mm for length of 3.8 cm located 3.4 cm from anal verge with enlarged mesorectal and extra-mesorectal lymph nodes (Figure 1). No distant metastasis was detected on CT scan. Patient was planned for abdominoperineal resection.

Intra-operatively, there was no ascites, liver metastasis or peritoneal/pelvic/omental deposits. A single polypoidal black colored growth of around 3 cm present around 1 cm from anal verge (Figure 2). It was involving posterior and lateral walls of anal canal. Urinary bladder, prostate, lateral pelvic wall and pre-sacral fascia were free. Multiple centimetric lymph nodes were present in mesorectum.

Surgery and post-operative period were uneventful. Patient had an uneventful recovery and was discharged on post-operative day (POD) eight.

Final histopathology revealed polypoidal tan-brown, partly ulcerated growth of 4 x 2.5 x 2 cm on cut-section. Microscopy showed sheets of round to oval spindle shaped tumor cells with high N:C ratio, fine chromatin, prominent nucleolus, moderate eosinophilic cytoplasm with markedly extensive extracellular and intracellular melanin pigment (Figure 3a,b and 4). These findings were consistent with malignant melanoma. All 15 lymph nodes dissected were free from tumor deposits.

#### Discussion and Conclusion

The incidence of anal canal melanoma increases with age with mean age of presentation being 60 years with female preponderance [12]. Most patients have extensive disease at presentation. Embryologically, melanocytes are derived from neural crest cells and present in anal canal and in transitional zone i.e., junction between squamous epithelium of anal canal and columnar epithelium of rectum [13]. Lesions can affect anal canal, rectum or both; but the great majority of tumours are located within 6 cm of the anal rim. Lesions can be melanotic or amelanotic (in 20 to 30% of cases) [14, 15].

Symptoms are generally non-specific, rectal bleeding being the commonest symptom. Diagnosis is even more difficult in amelanotic melanoma because they can resemble benign polypoid lesion on visual inspection [5, 6]. Diagnosis is based on per rectal examination and biopsy with immunohistochemical staining. EUS/ MRI with CT and/or PET imaging are helpful in

staging the disease. Histological examination characteristics of the lesions include cell type, degree of melanin pigmentation, mitotic index. On IHC, presence of protein S-100, melanoma antigen HMB-45, Melan-A and Mart-1 antibodies confirms the diagnosis [11].

Anorectal melanoma is excluded from the American Joint Committee on Cancer (AJCC) staging system for anal cancers. Retrospective series have used a simple system in which localized disease only, regional lymph node involvement, and distant metastases are classified as stages I, II, and III, respectively [16].

Most patients with distant metastases have hepatic metastases, followed by pulmonary and bone metastases. The most important prognostic indicators in anorectal melanomas include the stage of disease and nodal involvement. In patients with localized disease, presence of perineural invasion, tumour size and thickness, and the presence of amelanotic melanoma are indicative of poor prognosis [17].

The median survival of stage I is 24 months, stage II is 17 months and stage III is 8 months [5].

There is no unified consensus regarding treatment in anorectal melanoma due to absence of randomised trials. Various modalities are surgery, radiotherapy, chemotherapy, immunotherapy and targeted therapy [11].

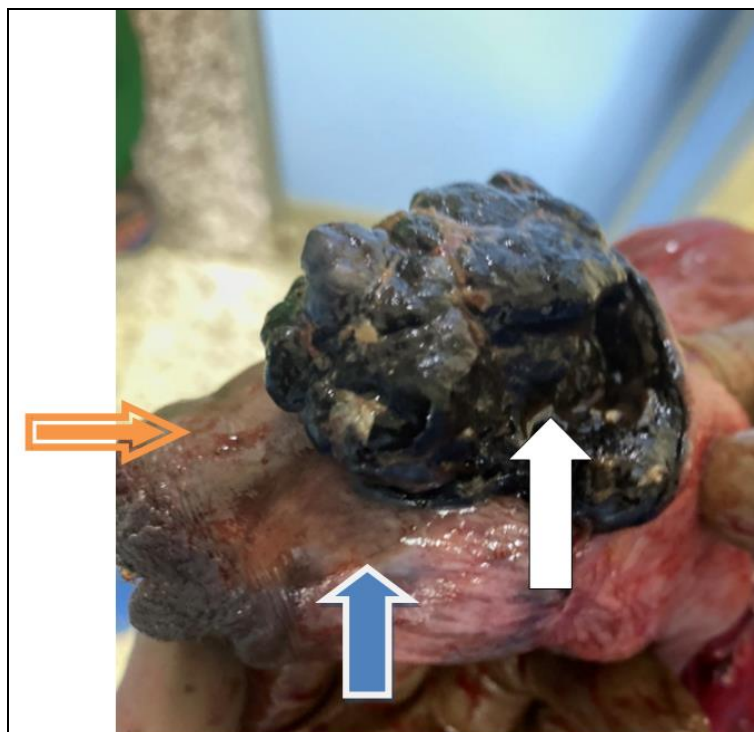
The typical treatment is surgical resection; however, standard operative procedures related to the area of resection and lymph dissection have yet to be established.

Traditionally, APR was considered best option for better loco-regional control but recent studies have challenged this notion. Now-a-days, WLE followed by adjuvant radiotherapy to the pelvis and inguinal lymph nodes employed in most of the patients due to the similar local control rate as APR, which is preferred in the cases with local extensive disease not amenable to a local excision [18].

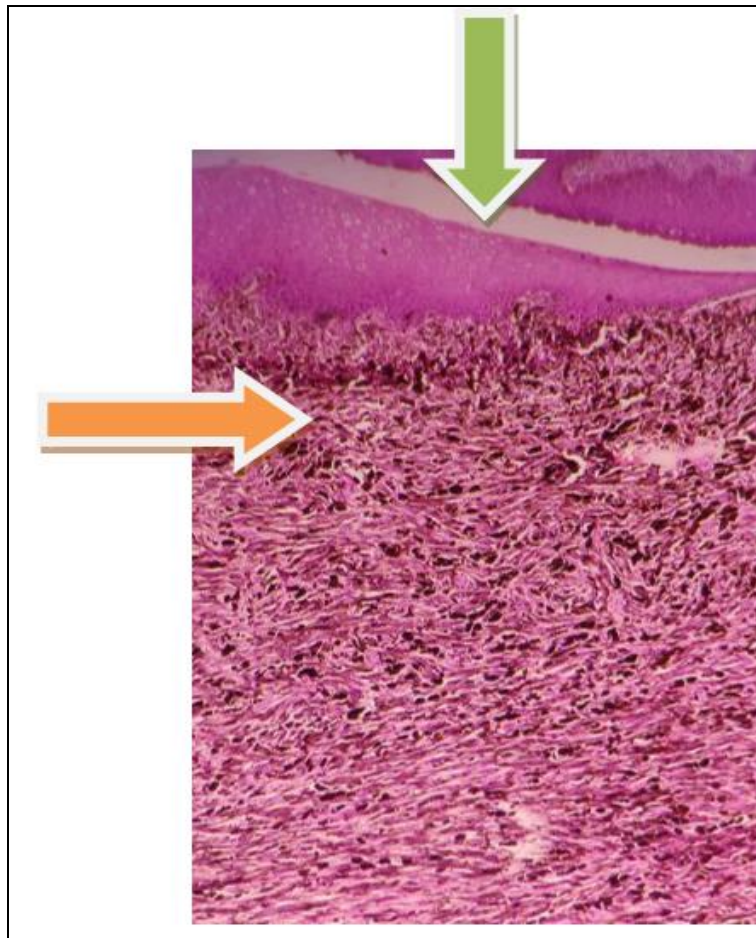
There are limited data on adjuvant systemic therapy in patients with mucosal melanoma. Dacarbazine is the most used commonly single agent and usually initiates a partial response in 20% of patients four to six months after treatment. In patients with positive nodal involvement and metastatic melanoma chemotherapeutic agents (Dacarbazine and Temozolomide) along with immunotherapy (alpha-Interferon and Interleukin-2) are used [19]. Newer treatment modalities including targeted therapies including c-KIT inhibitors, BRAF inhibitors, MEK-1/2 inhibitors, anti-CTLA-4 antibodies and anti PD-4 antibodies are used [20].



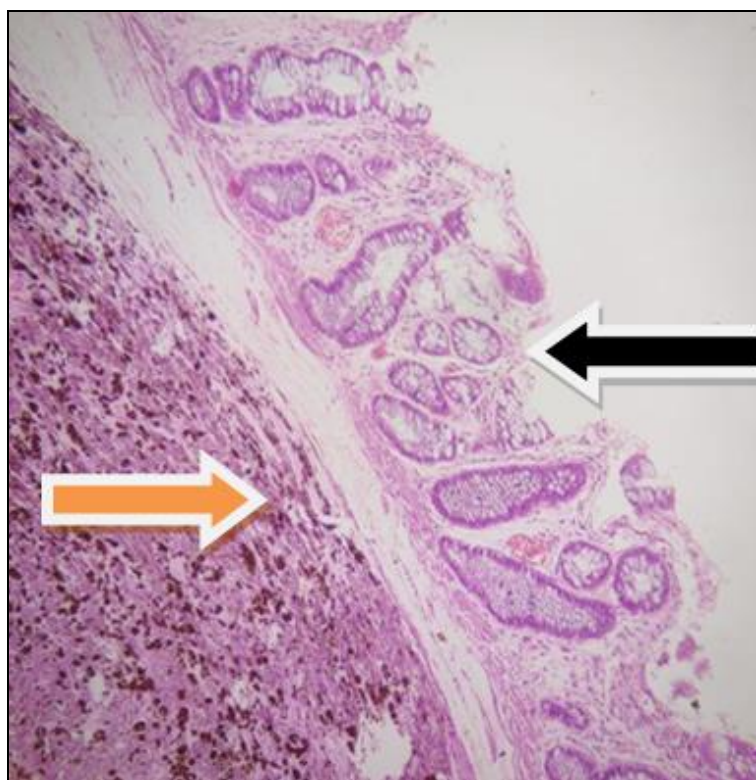
**Fig 1:** MRI showing asymmetrical circumferential thickening in anal canal region (red arrow)



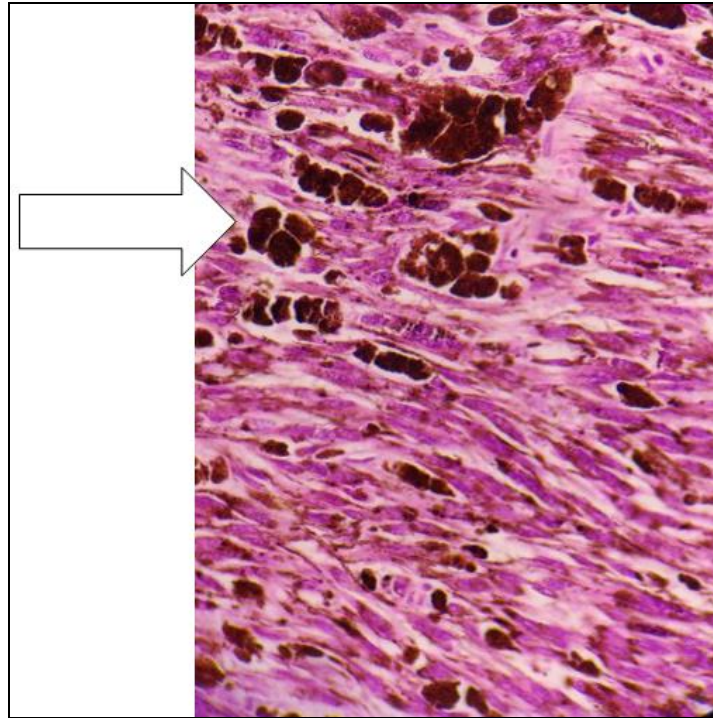
**Fig 2:** Resection specimen showing dentate line (orange arrow), anal verge (blue arrow) and ulcerated- pigmented polypoidal growth (white arrow)



**Fig 3a:** Microscopy showing anal squamous epithelium (green arrow) and melanin containing melanocytes (orange arrow)



**Fig 3b:** Microscopy showing rectal epithelial glands (black arrow) and melanin containing melanocytes (orange arrow)



**Fig 4:** Microscopy (100X) showing intra and extra-luminal melanin deposition (white arrow)

### Conclusion

Anorectal malignant melanoma is a rare form of anorectal malignancy. Biopsy along with IHC confirms the diagnosis. Treatment is surgical wide local excision for small tumors and abdominoperineal resection for large tumors with adjuvant chemoradiotherapy and immunotherapy. Targeted therapy and checkpoint inhibitors have an upcoming role. Despite all these treatment options, prognosis and overall survival is dismal.

**Acknowledgement:** None

**Funding:** None

**Conflict of Interest:** None

### References

- Cagir B, Whiteford MH, Topham A, *et al.* Changing epidemiology of anorectal melanoma. *Dis Colon Rectum* 1999;42:1203.
- Coté TR, Sobin LH. Primary melanomas of the esophagus and anorectum: epidemiologic comparison with melanoma of the skin. *Melanoma Res.* 2009;19:58.
- Burgi A, Brodine S, Wegner S, *et al.* Incidence and risk factors for the occurrence of non-AIDS-defining cancers among human immunodeficiency virus-infected individuals. *Cancer.* 2005;104:1505.
- Bello DM, Smyth E, Perez D, *et al.* Anal versus rectal melanoma: does site of origin predict outcome? *Dis Colon Rectum.* 2013;56:150.
- Iddings DM, Fleisig AJ, Chen SL, *et al.* Practice patterns and outcomes for anorectal melanoma in the USA, reviewing three decades of treatment: is more extensive surgical resection beneficial in all patients? *Ann Surg Oncol.* 2010;17:40.
- Pessaux P, Pocard M, Elias D, *et al.* Surgical management of primary anorectal melanoma. *Br J Surg.* 2004;91:1183.
- Cooper PH, Mills SE, Allen MS Jr. Malignant melanoma of the anus: report of 12 patients and analysis of 255 additional cases. *Dis Colon Rectum.* 1982;25:693.
- Weinstock MA. Epidemiology and prognosis of anorectal melanoma. *Gastroenterology.* 1993;104:174.
- Goldman S, Glimelius B, Pählman L. Anorectal malignant melanoma in Sweden. Report of 49 patients. *Dis Colon Rectum.* 1990;33:874.
- Ross M, Pezzi C, Pezzi T, *et al.* Patterns of failure in anorectal melanoma. A guide to surgical therapy. *Arch Surg.* 1990;125:313.
- Matsuda A, Miyashita M, Matsumoto S, *et al.* Abdominoperineal resection provides better local control but equivalent overall survival to local excision of anorectal malignant melanoma: a systematic review. *Ann Surg.* 2015;261:670.
- Zhong J, Zhou JN, Xu FP, Shang JQ. Diagnosis and treatment of anorectal malignant melanoma - a report of 22 cases with literature review. *Ai Zhong.* 2006;25:619-24.
- Sadler TW. (Ed.), *Langman's Med. Embryol.*, 14th ed., Wolters Kluwer, 2018, 230-255.
- Zhang S, Gao F, Wan D. Effect of misdiagnosis on the prognosis of anorectal malignant melanoma. *J Cancer Res Clin Oncol.* 2010;136:1401-05.
- Felz MW, Winburn GB, Kallab AM, Lee JR. Anal melanoma: an aggressive malignancy masquerading as hemorrhoids. *South Med J.* 2001;94:880-85.
- Singer M, Mutch MG. Anal melanoma. *Clin Colon Rectal Surg.* 2006;19:78.
- Prasad ML, Jungbluth AA, Patel SG, Iversen K, Hoshaw-Woodard S, Busam KJ. Expression and significance of cancer testis antigens in primary mucosal melanoma of the head and neck. *Head Neck.* 2004;26:1053-57.
- Kiran RP, Rottoli M, Pokala N, Fazio VW. Long-term outcomes after local excision and radical surgery for anal melanoma: data from a population database. *Dis Colon Rectum.* 2010;53:402-08.
- Wang X, Si L, Guo J. Treatment algorithm of metastatic mucosal melanoma. *Chin Clin Oncol.* 2014;3:38.
- Lutzky J. New therapeutic options in the medical management of advanced melanoma. *Semin Cutan Med Surg.* 2010;29:249-57.