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Case Report

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Anal Canal Melanoma - A Case Report and Review of Literature

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Abstract

Anorectal mucosal melanoma is one of the rarest neoplasms 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 abdomino-perineal 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 neoplasms and accounts for approximately 0.05 percent of all colorectal malignancies and 1 percent of all anal canal cancers 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 [1]. 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 [2, 3]. 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 [4]. Patients typically present with bleeding, a mass, anorectal pain, or a change in bowel habits [5, 6]. 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 [7]. Most patients present with lesions that are >2 mm thick [8]. 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 [9, 10].

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 abdomino-perineal resection depending upon the site, size and extent along with adjuvant chemotherapy, radiotherapy and immunotherapy [11]. 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).

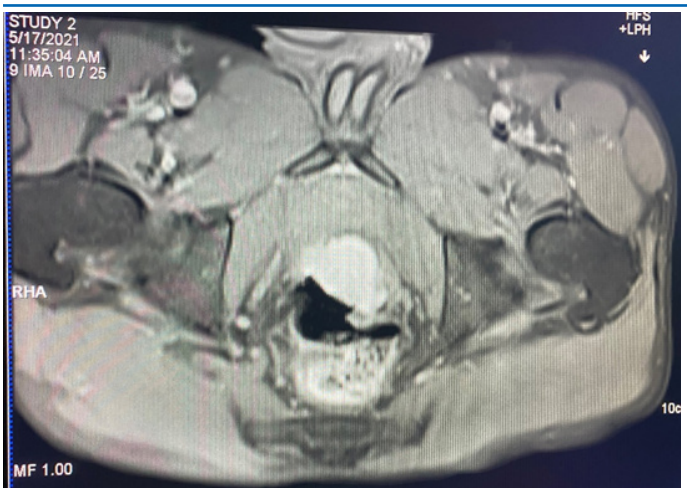

Figure 1: MRI showing asymmetrical circumferential thickening in anal canal region (red arrow)

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).

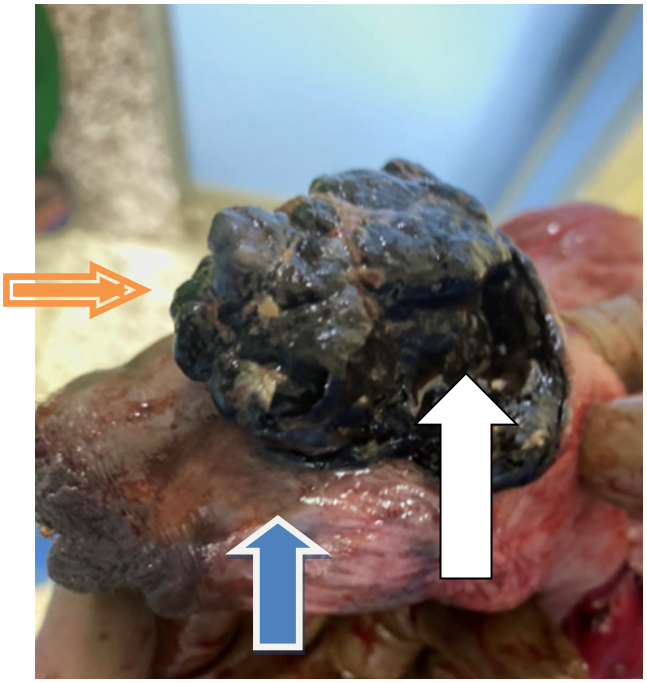

Figure 2: Resection specimen showing dentate line (orange arrow), anal verge (blue arrow) and ulcerated- pigmented polypoidal growth (white arrow)

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).

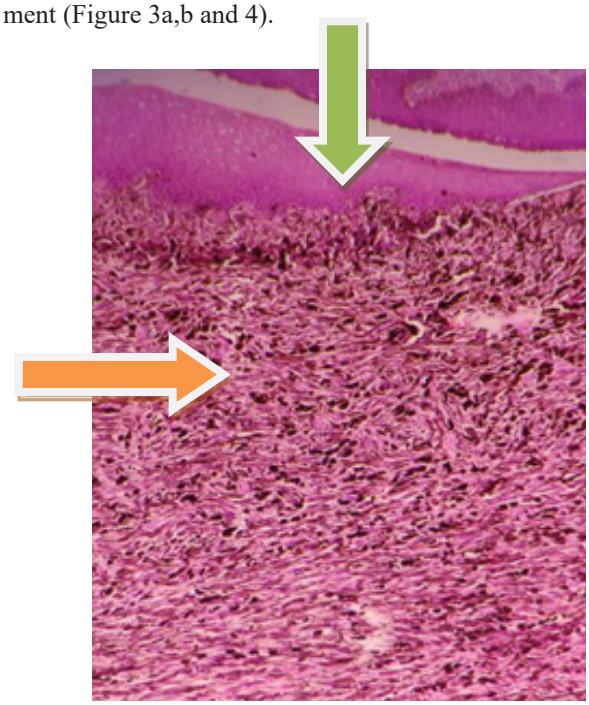

Figure 3a: Microscopy showing anal squamous epithelium (green arrow) and melanin containing melanocytes (orange arrow)

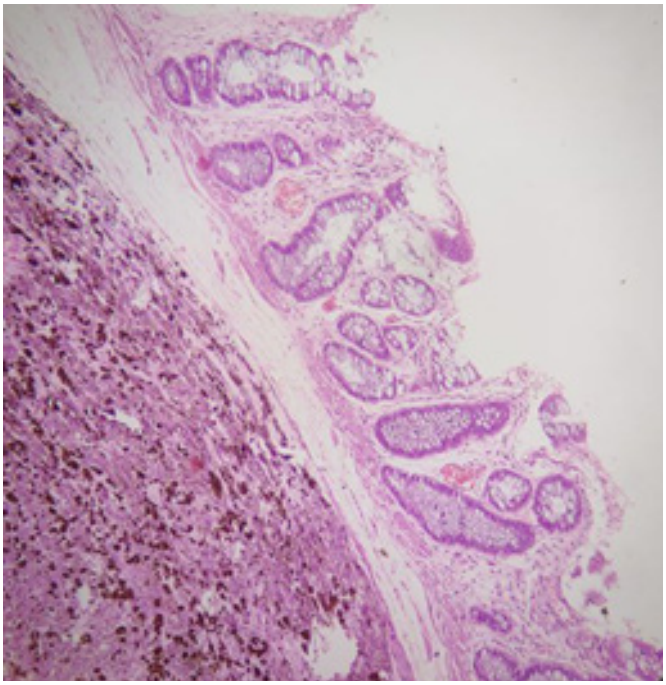

Figure 3b: Microscopy showing rectal epithelial glands (black arrow) and melanin containing melanocytes (orange arrow)

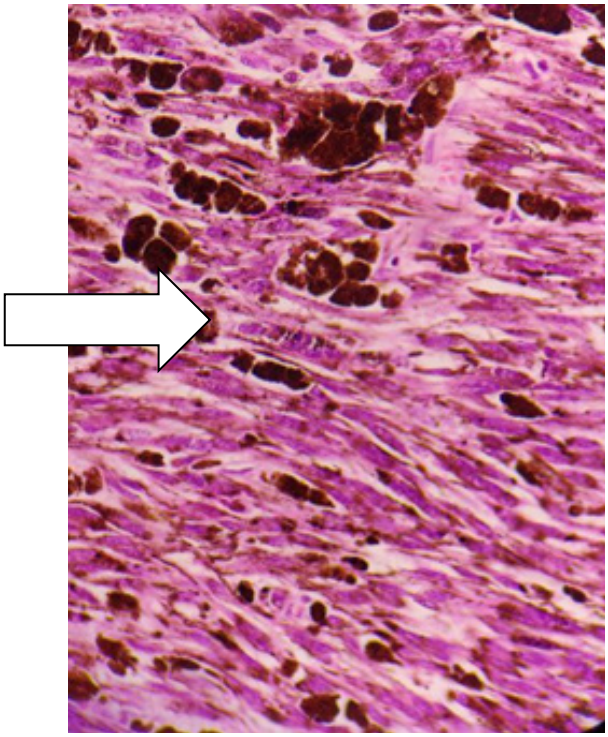

Figure 4: Microscopy (100X) showing intra and extra-luminal melanin deposition (white arrow)

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. 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 characterics 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. 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 is17 months and stage III is 8 months. 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. 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].

Conclusion

Anorectal malignant melanoma is a rare form of anorectal malignancy. Biopsy along with IHC confirms the diagnosis. Treatment is surical 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.

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