Metastatic Malignant Melanoma of Rectum – A few and far between uncommonness

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ABSTRACT

INTRODUCTION: Malignant melanoma of rectum is an extremely rare and very aggressive neoplasm. It accounts for only 1% of anorectal malignancies and less than 1% of all melanomas. Patients often present with nonspecific complaints such as rectal bleeding, anal pain or change in bowel habit. Prognosis of malignant melanoma of rectum is very poor with a median survival of 24 months and a 5-year survival of 10-15%.

CASE PRESENTATION: A 59-year-old male presented with a 10-month history of bleeding per rectum. MRI abdomen and pelvis revealed a 22 x 21 x 18 mm heterogeneously enhancing lesion over the right anterolateral wall of ano-rectum. Multiple enlarged mesorectal lymph nodes were seen. Right kidney showed two well-defined exophytic lesions of 15 x 12 mm and 12 x 9 mm and a well-defined soft tissue lesion of 36 x 26 x 25 mm in right suprarenal region. Patient underwent rectal polypectomy and the histopathology revealed malignant melanoma. On immunohistochemistry, S-100, HMB-45 and melan-A were positive. For adjuvant treatment, the patient was planned to receive dacarbazine and tamoxifen based chemotherapy.

CONCLUSION: Malignant melanoma of rectum is an extremely rare and very aggressive neoplasm. Surgery remains the mainstay of treatment for limited disease. There are no standard solidarity recommendations for systemic therapy for disseminated disease. More research protocols & clinical trials should be explored to improvise survival in these patients.

INTRODUCTION

Malignant melanoma of rectum is an extremely rare and very aggressive neoplasm.[1] It accounts for only 1% of anorectal malignancies and less than 1% of all melanomas.[2,3] It is even more rare in males and typically occurs in the fifth or sixth decade of life. Unlike other forms, there is no association with exposure

to ultraviolet light. Malignant melanoma is strongly associated with the Caucasian race. Cutaneous melanoma is 20 times more common in Caucasians than in African Americans, but there is no such evidence for anorectal melanomas.

Patients often present with nonspecific complaints such as rectal bleeding, anal pain or change in bowel habit.[1-3] Prognosis of malignant melanoma of rectum is very poor with a median survival of 24 months and a 5-year survival of 10-15%.[1,6] Presently, there is no consensus that supports surgical approach favorable.[3] Anorectal malignant melanomas spread along submucosal planes and thus, they are often beyond complete resection at the time of diagnosis[7] and almost all patients die because of metastases.[8]

Here we present a case of malignant melanoma of rectum and discuss the management done.

CASE PRESENTATION

A 59-year-old male presented with a 10-month history of bleeding per rectum. General physical examination and systemic examination were normal. Local examination of abdomen revealed scaphoid abdomen with no tenderness or organomegaly. No growth was palpable on per rectal digital examination. Hematological and biochemical profile were within normal limits. Chest X-ray was also normal.

MRI abdomen and pelvis revealed a 22 x 21 x 18 mm heterogeneously enhancing lesion over the right anterolateral wall of ano-rectum with infiltration into the pelvic floor muscles. Multiple enlarged mesorectal lymph nodes were seen (largest measuring 7 x 6.5 mm). Right kidney showed two well-defined exophytic lesions of 15 x 12 mm and 12 x 9 mm and a well-defined soft tissue lesion of 36 x 26 x 25 mm in right suprarenal region. Patient underwent rectal polypectomy and the histopathology revealed malignant melanoma. On immunohistochemistry, S-100, HMB-45 and melan-A were positive.

For adjuvant treatment, the patient was planned to receive dacarbazine and tamoxifen based chemotherapy. The patient received 1 course of chemotherapy and thereafter the general condition of the patient deteriorated and patient was not able to tolerate further courses of chemotherapy. The patient is presently on follow up for 2-months and is on best supportive care.

DISCUSSION

Malignant melanoma of rectum is an extremely rare and very aggressive neoplasm.[1] It accounts for only 1% of anorectal malignancies and less than 1% of all melanomas.[2,3] Melanomas originate from melanocytes, which are the cells derived from the embryological neural crest. During embryonal development, these cells migrate to different sites in the body, residing in the skin, mucous membrane, meninges and eyes.[2,3] Melanocytes undergo malignant transformation when they are exposed to ultraviolet radiation. Ultraviolet light acts as a carcinogenic agent. However, this relationship is not discernible in anorectal melanoma.[7] There may be a role of immune system in the evolution of anorectal melanoma as the incidence is higher in patients with human papilloma virus (HPV) and HIV infections.[7,9] In rectum, melanocytes are present in anal transition zone and squamous epithelial zone. The most common presenting features are bleeding per rectum, anorectal discomfort or pain, anorectal mass, alteration in bowel habits, pruritis, tenesmus, prolapsed hemorrhoid, and diarrhea.[3,7] Sigmoidoscopy or colonoscopy shall be performed for evaluation of the cause of symptoms and obtaining a tissue biopsy from a suspicious lesion. Endoscopic ultrasound of anorectum can also be considered to evaluate tumor thickness and surrounding nodal status.[7] MRI shows melanotic component as high signal intensity on T1-weighted imaging and mixed signal intensity on T2-weighted imaging.[10] Extraluminal extent of the lesion is better visualized by MRI imaging. Other rectal mass lesions show hypointense signal on T1-weighted imaging. Melanoma antigens S-100, HMB-45, and melan-A are important immuno-histochemical markers.

The treatment of anorectal melanoma, unfortunately, is only moderately successful. Surgery remains the mainstay of treatment.[1,2] The 5-year survival rate of anorectal melanoma ranges from 16 to 34%. In patients who have metastasis at the time of diagnosis, the disease-free survival rate may drop to 16% from 22%. There are no standard solidarity recommendations for systemic therapy for disseminated disease. Chemotherapy, radiation therapy, and immune therapy have a limited role. The medications used in adjuvant therapy are cisplatin, vinblastine, dacarbazine, interferon B, and Interleukins- 2-8. Dacarbazine is the most commonly used single agent and gives a partial response in 20% of patients in 4-6 months after treatment.[2,7]

CONCLUSION

Malignant melanoma of rectum is an extremely rare and very aggressive neoplasm. Surgery remains the mainstay of treatment for limited disease. There are no standard solidarity recommendations for systemic therapy for disseminated disease. More research protocols & clinical trials should be explored to improvise survival in these patients.

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