Malignant Melanoma of the Anorectum: A Bizarre Presentation

Amit Gupta1, Amulya Rattan2, Manish Kumar3, Lovenish Bains4, Chander Mohan5, Neelam Wadhwa6

1associate Professor Dept Of Surgery Dr Baba Saheb Ambedkar Medical College Hospital, 2post Graduate Dept Of Surgery, Ucms & Gtb Hospital, 3assistant Professor Dept Of Surgery Dr Baba Saheb Ambedkar Medical College Hospital, 4senior Resident Dept Of Surgery, 5associate Professor Dept Of Pathology, Ucms & Gtb Hospital, New Delhi

ABSTRACT

Anorectal melanoma is a rare malignancy accounting for only 1-2 % of anorectal tumors. The absence of early clinical manifestations and the lack of clinical suspicion contributes for delayed diagnosis. Owing to its rarity and histologic variability, misdiagnosis as lymphoma, carcinoma, or sarcoma is common. The propensity of MM to metastasize early, diffusely and unpredictably is responsible for invariably disappointing outcomes.

Key words: Anorectal, Melanoma, Surgery

Corresponding author address: Dr Amit Gupta, B-1 503,Olive County,sector-5,Vasundhara,Ghaziabad (U.P.) India. Pin code – 201012 E-Mail: dramit2411@yahoo.co.in M: 9891981331

Conflict of interest: No

Case report is Original: YES

Whether case report publishes any where? NO

INTRODUCTION

Malignant melanoma(MM) is a rare tumor of the anorectum, accounting for only 1% of the cases at this site. It is mainly seen in sixth decade of life and male to female ratio is 1:1. At the time of presentation, 20% of patients have regional and 40% have distant metastases. Metastasis to nodes, lungs, liver, bone and brain are known to occur. Mucosal melanomas usually have a poor prognosis.. Treatment options are equivocal due to scarcity of clinical trials. For a localized disease, opinions vary from aggressive approach to local therapies, but the outcome remains miserable in spite of multimodal therapy. Adjuvant radiation may decrease locoregional recurrence but overall survival remains poor.

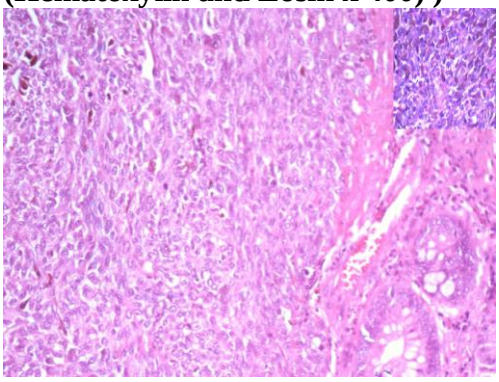
CASE REPORT

A 55 year old lady presented to surgical outpatient department with complaints of mass protruding through anus for 6 months which was gradually progressive and recently became painful . It was associated with bleeding per rectum and constipation. On examination a grossly infiltrating growth per anum with luminal compromise was seen. Whole of the anal canal was distorted, a large growth with induration in the anterior wall of the anal canal reaching up to the lower rectum. the full extent of the growth could not be assessed on digital

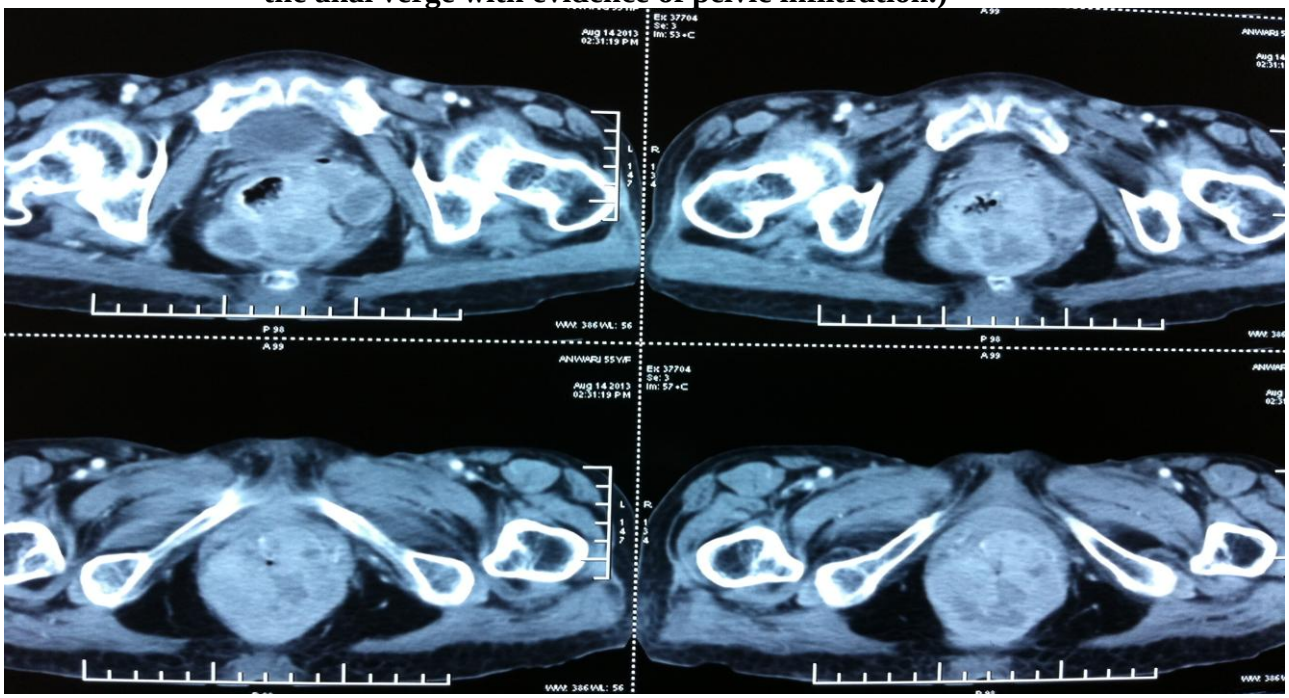
rectal examination (Figure:1). On per vaginum examination, a 7 cm long growth was felt in the anterior wall of rectum through the posterior wall of vagina. Both fornices were free and vaginal mucosa was mobile. Punch biopsy from the growth revealed malignant melanoma(Figure:2). Contrast enhanced CT abdomen showed a long luminal growth from rectosigmoid junction to the anal verge with evidence of pelvic infiltration (Figure:3). Diversion colostomy was done in view of advanced nature of disease with features of partial large bowel obstruction and patient was referred to medical oncology for palliative chemotherapy.

(Fig: 1 Clinical photograph showing pigmented lesion involving anal verge)

(Figure 2– Malignant melanoma composed of tumor cells with prominent nucleoli. (Hematoxylin and Eosin x 200) Figure Inset – Abundant pigment in tumor cells, from another area. (Hematoxylin and Eosin x 400))



(Fig 3: CECT pelvis showed luminal growth extending from rectosigmoid junction to the anal verge with evidence of pelvic infiltration.)



DISCUSSION

Anorectal melanoma is a rare malignancy accounting for only 1-2 % of anorectal tumors.[1] The anorectum is the third most common location of malignant melanoma after the skin and retina. Malignant melanomas occur commonly in the anorectum because of the presence of abundant melanocytes in the mucosa of the anal canal. The metastases are highly unpredictable and skip lesions are possible. Upto 20% patients have regional disease at presentation and 40% present with distant metastasis to nodes, lung, liver, bone and brain. The absence of early clinical manifestations and the lack of clinical suspicion contributes for delayed diagnosis. Owing to its rarity and histologic variability, misdiagnosis as lymphoma, carcinoma, or sarcoma is common[2,3]. Histopathological examination and immunohistochemical studies are of great importance in the diagnosis of anorectal MM. Positive protein S-100, melanoma antigen HMB- 45, and melan-A expression strongly support the diagnosis of melanoma[4]. Anti–S-100 protein is the most common screening immunohistochemical stain used in the diagnosis of malignant melanoma and is highly sensitive for melanocytic differentiation[5]. The main elements of prognosis are the depth of invasion and stage of the disease[6]. The factors for poor prognosis include, advanced disease at the time of diagnosis and rich vascularity which increases the risk of hematogenous metastasis[7]. Abdomino-perineal resection (APR) is the treatment of choice for patients with <2 mm wide lesion[8]. Radiotherapy is palliative in locally extensive tumors while combined with chemotherapy is used for metastasis. Chemotherapy is normally used for palliative purposes in advanced stages of MM and survival after diagnosis is quite short. There are standard systematic treatment options defined in advanced cutaneous MM patients, including cisplatin, vinblastine, DTIC, IFN, and interleukin-2. Because of the limited number of studies, there is no standard treatment for mucosal MM. Another alternative is an orally bioavailable drug, temozolamide. Yeh et al.[9] used a combination regimen with cisplatin as the third line of treatment after colostomy and radiotherapy, TMZ, and liposomal doxorubicin for a 49-year-old female anal mucosal melanoma patient with complete colonic obstruction and multiple distant organ metastases. After the second course, more than 50% regression was observed in the metastases in all regions, Ipilimumab, which is an immunomodulatory monoclonal antibody, was developed against an antigen on T lymphocytes. MM treatment should be further studied. The prognosis is very poor, with less than 20% survival five years after diagnosis.

CONCLUSION

Malignant melanoma of the anorectum is a rare disease and high volume centers are expected to come across only one case per year. Due to the rarity of this disease, the pathophysiology remains poorly understood. The propensity of MM to metastasize early, diffusely and unpredictably is responsible for invariably disappointing outcomes. Clinicians should suspect anorectal melanoma in cases presenting with blood in the stool. Furthermore, the prognosis depends on the staging, and it is important to detect anorectal melanoma at an early stage. Presently, advanced anorectal MM remains an incurable disease, and despite the use of multidisciplinary strategies (radical surgery, immunotherapy, chemotherapy, and radiotherapy), it remains a fatal disease.

REFERENCES

1. Wanebo HJ, Woodruff JM, Farr GH, Quan SH. Anorectal melanoma. Cancer 1981; 47: 1891–900
2. Kuroda T, Kusama J, Iijima K, et al. Primary malignant melanoma of the rectum. J Gastroenterol. 1996;31:437-440.
3. Row D, Weiser MR. Anorectal melanoma. Clin Colon Rectal Surg 2009; 22: 120–6.
4. Deborah J. Chute, John B. Cousar, Stacey E. Mills. Anorectal Malignant Melanoma: Morphologic and Immunohistochemical Features. Am J Clin Pathol: 2006; 126:93-100
5. Vijayaraghavan M, Kharbanda K, Mathur M. S-100 staining for the diagnosis of melanoma of the anal canal. Indian J Pathol Microbiol. 1992;35:113-117.
6. Antoniuk PM, Tjandra JJ, Webb BW, Petras RE, MilsonJW, Fazio VW. Anorectal malignant melanoma has a poor prognosis. Int J Colorectal Dis 1993; 8: 81–6.
7. Carlson JA, Slominski A, Linette GP, et al. Malignant melanoma 2003: predisposition, diagnosis, prognosis, and staging. Am J Clin Pathol. 2003;120(suppl):101-S127
8. Pessaux P, Pocard M, Elias D, Duvillard P, Avril MF, Zimmerman P et al. Surgical management of primary anorectal melanoma. Br J Surg 2004; 91: 1183–7.
9. Yeh JJ, Weiser MR, Shia J, Hwu WJ. Response of stage IV anal mucosal melanoma to chemotherapy. Lancet Oncol 2005; 6:438–9.