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Malignant Melanoma of the Anal Canal - A Rare Case Report from a Tertiary Care Center in South India

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ABSTRACT

Anorectal melanoma is a rare entity that accounts for less than 1% of all anorectal malignancies. It is often mistaken for benign anorectal conditions since the symptoms resemble each other. The predominant symptoms of anorectal melanoma are per rectal bleeding, anorectal pain, mass per rectum, tenesmus and altered bowel habits. All suspected cases of anorectal malignancy should be subjected to sigmoido-colonoscopy and biopsy. Management of anorectal melanoma remains controversial including surgery, chemotherapy, radiotherapy and target therapy as treatment options. Surgical treatment is considered as the primary treatment modality for anorectal melanoma. Anorectal melanoma must be kept in mind as differential diagnosis for patients pesenting with per rectal bleeding and pain.

Keywords: Melanoma, Anorectum, Carcinoma, Abdominal perineal resection

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INTRODUCTION

Anorectal melanoma is a rare entity that accounts for less than 1% of all anorectal malignancies.[1,2] It is often mistaken for benign anorectal conditions since the symptoms resemble each other.[3] The predominant symptoms of anorectal melanoma are per rectal bleeding, mass per rectum, and altered bowel habits.[4] All suspected cases of anorectal malignancy should undergo sigmoido-colonoscopy and biopsy. Management of anorectal melanoma remains controversial including surgery, chemotherapy, radiotherapy and target therapy as treatment options. Surgical treatment is considered as the primary treatment modality for anorectal melanoma. We present the case of a patient who presented to the outpatient department of a tertiary care centre in south India.

CASE REPORT

61 year old lady presented with complaints of progressive constipation for the past 8 months. She gave history of passage of blood and mucus in stools and intermittent lower abdominal pain of 1 year duration. She complained of fatigue and significant weight loss in the past 6 months. On admission pulse rate was 80 beats per minute, blood pressure 130/80mm Hg. On examination pallor was present. Abdomen was soft and non distended without any palpable mass. Per rectal examination revealed a large, fleshy, firm mass along the six o clock to twelve o clock position in the distal rectum and anal canal. The lower limit of the mass was less than 1 cm from the anal verge and the proximal limit nearly 5cm from the anal verge. There was no blood staining. The anal sphincter was intact. There were no palpable inguinal lymph nodes. She was suspected to have malignancy involving the rectum and anal canal and evaluated with USG abdomen and CEMRI pelvis. CEMRI pelvis revealed a polypoidal lobulated mass measuring 4.6cm X 4.0cm X 7.4cm (AP X TR X CC) within the lumen of the rectum with the stalk being 4.6cm from the anal verge. Effacement of the rectal walls was noted. Significant invasion of the submucosa and muscularis propia at 9 o clock position was noted. Two lymph nodes measuring 0.5cm X 0.cm were seen in the right pararectal fossa with normal enhancement. Visceral pelvic fat, anal sphincters and pararectal fat was normal. USG abdomen was done to rule out liver

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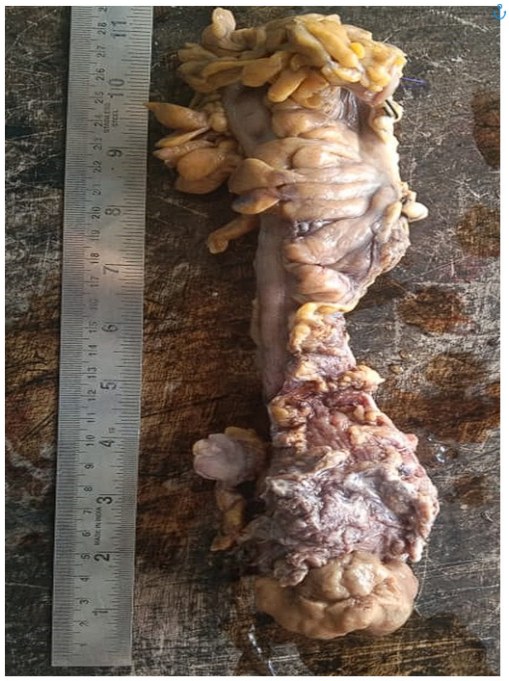

Figure 1. **Colonoscopy images showing tumor at anorectal region**

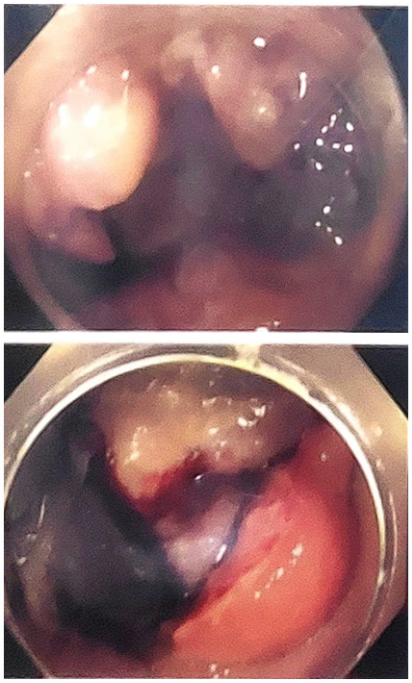

Figure 2. **Abdominoperineal resection specimen showing tumor at inferior end**

metastasis and ascites and it was reported as normal. She underwent colonoscopy **(Figure 1)** which revealed an ulceroproliferative mass extending from the anal canal into the rectum, likely to be malignant. Colonoscopic/colonoscopy guided biopsy was also taken. The biopsy was reported as spindle cell tumor/ gastrointestinal tumor (GIST) of the anal canal. IHC was strongly positive for CD117, suggestive of GIST. She was managed in consultation with Medical oncology team and planned for abdominal perineal resection. She underwent abdominal perineal resection **(Figure 2)** under general anesthesia. Intraoperatively the rectosigmoid was mobilised and descending colon divided midway via midline laparotomy approach. The rectum and anal canal were dissected via perineal approach. A highly friable fleshy growth arising from just above the posterior anal margin and extending cranially for a length of 7cm in contact with the right lateral wall was noted. The posterior vaginal wall was found in close contact with tumour. The descending colon, rectum and anal canal was delivered via the perineal wound. End colostomy was fashioned in the left iliac fossa. Postoperatively she was managed in the surgical ICU and subsequently in the high dependency unit with intravenous antibiotics, analgesics, and other supportive measures. She was started on oral diet on postoperative day 2 and colostomy was functional within 48 hours of surgery. Her condition gradually improved and she was discharged on post operative day ten after suture removal. Histopathology of the resected specimen showed 5cm X 4.5cm X 4cm lesion with spindle cells, coarse chromatin, irregular nuclear membrane and moderate cytoplasm. Brownish coarse pigment consistent with melanin was noted in occasional tumour cells. Surgical margins were uninvolved. 2 lymph nodes identified were uninvolved. Perineural and angiolymphatic invasion were absent. Immunohistochemistry was positive for CD 117 and S100 suggestive of spindle cell type of malignant melanoma of the anal canal. She was followed up on outpatient basis and surgical wounds had healed well. She has been advised PET scan to rule out metastases and follow up after 3 months.

DISCUSSION

The hindgut develops into the distal part of the transverse colon, descending colon, rectum and anal canal. The dentate line divides the anal canal into an upper and lower part and marks the junction between endoderm and ectoderm. At the dentate line the columnar epithelium of the upper anal canal changes into stratified squamous epithelium. Melanocytes are located in the squamous zone. Melanocytes originate from the neural crest and migrate towards the skin and other regions.[5,6]

The first case of malignant melanoma of the anorectum was reported by Moore in 1857.Since then nearly 500 cases have been reported till date.[7]

Cutaneous melanomas account for nearly 90% of all melanomas followed by retinal melanomas (5%), melanomas of unknown origin (2%) and mucosal melanomas(1%).[8] Mucosal melanomas occur in the

regions of head and neck(55%), anorectum (24%) and vulvovaginal areas (18%).[9] Anorectal melanomas are extremely rare and usually arise near the dentate line and can rarely be found in the distal ileum. They account for 0.5%-4% of all anorectal malignancies. It is the third most common primary melanoma after skin and retina.

The mean age of presentation is in the sixth decade of life.[10] It is more common in females than males.[11]

The predominant symptoms of anorectal melanoma are per rectal bleeding, anorectal pain, mass per rectum, tenesmus and altered bowel habits.[4] Pain occurs since most of the melanomas are located at or near the anal verge which is rich in nerve fibres. The most common presentation is per rectal bleeding. Most patients are asymptomatic in the early stages and especially more so in cases of amelanotic melanoma. Anorectal melanomas are frequently mistaken for hemorrhoids and other benign anorectal diseases due to its rarity.[3] Amelanotic melanomas maybe mistaken for lymphoma or sarcoma.[12] This leads to diagnostic dilemma and delay in the diagnosis.

All suspected cases of anorectal malignancy should undergo sigmoido-colonoscopy and biopsy to confirm the diagnosis and rule out any synchronous or metachronous lesions in the colon or anorectum.[13] Majority of the lesions are polypoid. They maybe ulcerated as well.[14] One third of anorectal melanomas are amelanotic.[12] CECT and MRI scan of the abdomen & pelvis help in staging the disease particularly if chemotherapy or radiation therapy are considered in the treatment.[15] Staging of anorectal melanomas differs from that of cutaneous melanomas (TNM STAGING).

PET scan has been used for staging cutaneous melanoma and maybe useful for staging anorectal melanomas as well.[16]

Immunohistochemistry plays a significant role in the diagnosis of anorectal melanomas. S-100 protein, HMB -45, Melan-A and tyrosinase are the commonly used stains positive for melanomas. Among these, melan A and HMB-45 are highly specific.[17-21]

Management of anorectal melanoma remains controversial including surgery, chemotherapy, radiotherapy and target therapy as treatment options. Surgical treatment is the primary treatment modality. However, standard operative procedures have not been established. Abdominoperineal resection was thought to be the standard treatment[22,23] with wide local excision gaining more attention.[23-25] Abdominoperineal resection offers the benefit of safe resection margins and lymphatic control. Wide local excision has advantages like less invasive procedure, faster recovery and absence of colostomy.

A study by Brady et al[22] in 1995 suggested that aggressive treatment with abdominoperineal resection had favourable outcomes. In recent years there has been a shift towards less radical procedures. According to the Yeh et al[26] study, there is no significant difference between patients treated with abdominoperineal resection and wide local excision.

Endoscopic mucosal resection has been performed by some surgeons with long term survival (>6years) achieved in several cases.[27]

Ramakrishnan et al showed that survival rates were better in patients who received wide local excision with radiotherapy when compared to patients who underwent surgery alone.

There has been no proven benefit of inguinal lymphadenectomy. Lymph node dissection maybe indicated in clinically apparent disease or for occult disease identified with sentinel lymph node.

Patients are often diagnosed at the late stages and require abdominoperineal resection as definitive or palliative procedure as in our case.

Anorectal melanoma has a very dismal prognosis with a 10-19 months survival after diagnosis.[22] One year survival rate is 21% and five year survival rate is 37%.

CONCLUSION

Malignant melanoma of the anorectum is extremely rare and accounts for less than 1% of all anorectal malignancies. It is often diagnosed in the late stages as it is easily mistaken for benign conditions of the anorectum. Anorectal melanoma should be kept in mind as differential diagnosis for patients presenting with per rectal bleeding and pain. Surgery is the mainstay of treatment with abdominoperoneal resection and wide local excision being commonly performed procedures for anorectal melanoma. Early diagnosis and radical surgery are key factors in prompt management of anorectal melanoma.

END NOTE

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REFERENCES

4. Klas JV, Rothenberger DA, Wong WD, Madoff RD. Malignant tumors of the anal canal: the spectrum of disease, treatment, and outcomes. Cancer. 1999;85(8):1686-93.
5. Roumen RMH. Anorectal melanoma in the Netherlands: a report of 63 patients. Eur J Surg Oncol. 1996;22(6):598-601
6. Wanebo HJ, Woodruff JM, Farr GH, Quan SH. Anorectal melanoma. Cancer. 1981;47:1891-900.
7. Heeney A, Mulsow J, Hyland JM. Treatment and outcomes of anorectal melanoma. Surgeon. 2011;9:27.
8. Malaguarnera G, Madeddu R, Catania VE, Bertino G, Morelli L, Perrotta RE, et al. Anorectal mucosal melanoma. Oncotarget. 2018 Feb 2;9(9):8785–800.
9. T.W. Sadler, Digestive system, in: T.W. Sadler (Ed.), Langman's Med. Embryol., 14th ed., Wolters Kluwer, 2018, pp. 230–255.
10. Podnos YD, Tsai NC, Smith D. Factors affecting survival in patients with anal melanoma. Am Surg. 2006;72:917.
11. Chang AE, Karnell LH, Menck HR. The National Cancer Data Base report on cutaneous and noncutaneous melanoma: a summary of 84,836 cases from the past decade. The American College of Surgeons Commission on Cancer and the American Cancer Society. Cancer 1998;83:1664-78.
12. Anal melanoma. Singer M, Mutch MG. Clin Colon Rectal Surg. 2006;19:78–87.
13. Iddings DM, Fleisig AJ, Chen SL, Faries MB, Morton DL. 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;17:40-4.
14. Ragnarsson-Olding BK, Nilsson PJ, Olding LB, Nilsson BR. Primary ano-rectal malignant melanomas within a population-based national patient series in Sweden during 40 years. Acta Oncol 2009; 48:125-31.
15. Banerjee SS, Harris M. Morphological and immunophenotypic variations in malignant melanoma. Histopathology 2000;36:387- 402.
16. Anorectal melanoma. Stefanou A, Nalamati SP. Clin Colon Rectal Surg. 2011;24:171–176.
17. Chute DJ, Cousar JB, Mills SE. Anorectal malignant melanoma: morphologic and immunohistochemical features. Am J Clin Pathol 2006;126:93-100.
18. 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;232:181-6.
19. Prichard RS, Hill AD, Skehan SJ, O'Higgins NJ. Positron emission tomography for staging and management of malignant melanoma. Br J Surg 2002;89:389-96.
20. Y.C. Mishra, N. Nadkarni, A.V. Mainker, Primary oral malignant melanoma: a case report, Ann. Dent. 48 (1989) 12–15.
21. Chute DJ, Cousar JB, Mills SE. Anorectal malignant melanoma: morphologic and immunohistochemical features. Am J Clin Pathol. 2006;126:93-100.
22. Cruz GMG da, Andrade Filho J de S, Patrus G, Leite SM de O, Silva IG da, Teixeira RG, et al. Anorectal melanoma - histopathological and immunohistochemical features and treatment. J Coloproctol (Rio J). 2014 Jun;34:95–103.
23. Tomicic J, Wanebo HJ. Mucosal melanomas. Surg Clin North Am. 2003;83:237–52.
24. Somran J, Kanngurn S, Porncharoenpong S, Lertkajornsin O. Anorectal malignant melanoma: report of two cases from Buddhachinnaraj Hospital. J Med Assoc Thai. 2005;88:1128–33
25. Brady MS, Kavolius JP, Quan SH. Anorectal melanoma. A 64-year experience at memorial sloan-kettering cancer center. Dis Colon Rectum. 1995;38:146–51.
26. Yap LB, Neary P. A comparison of wide local excision with abdominoperineal resection in anorectal melanoma. Melanoma Res. 2004;14:147–50
27. Droesch JT, Flum DR, Mann GN. Wide local excision or abdominoperineal resection as the initial treatment for anorectal melanoma? Am J Surg. 2005;189:446–9.
28. Terada R, Ito S, Kobayashi M, Akama F, Tsujimura M, Ooe H. Anorectal melanoma: successful treatment by surgical excision and combination chemoimmunotherapy. Hepatogastroenterology. 2002;49:1545–8
29. Yeh JJ, Shia J, Hwu WJ, Busam KJ, Paty PB, Guillem JG, Coit DG, Wong WD, Weiser MR. The role of abdominoperineal resection as surgical therapy for anorectal melanoma. Ann Surg. 2006;244:1012–7.
30. J.H. Park, J.R. Lee, H.S. Yoon, T.Y. Jung, E.J. Lee, J.G. Lim, S.Y. Ko, J.H. Wang, J.D. Lee, H.Y. Kim, Primary anorectal malignant melanoma treated with endoscopic mucosal resection, Intest. Res. 13 (2015) 170.