



PRIMARY ANAL MALIGNANT MELANOMA: A RARE CASE REPORT

General Surgery

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ABSTRACT

Primary anal melanoma is rare and aggressive malignancy. The disease is undetected or mistaken for a benign polyp or haemorrhoids thus patients commonly present with advanced, even metastatic disease. Unlike cutaneous melanoma, anal melanoma has no known risk factors. Extracutaneous melanomas require special consideration due to their late diagnosis and consequently poor prognosis. Extracutaneous melanomas are considered to be biologically more aggressive than their cutaneous counterparts. Surgical excision remains the cornerstone of therapy. This case is reported because of the rarity of the disease in anal canal found in an 65 year old male who was managed with diversion sigmoid loop colostomy: to relieve obstruction.

KEYWORDS

melanoma, malignancy, anal canal

INTRODUCTION:

Anorectal melanoma is an uncommon and aggressive cancer with an unfavorable prognosis¹⁻⁴ and a predilection for early infiltration and distant spread, resulting in poor overall survival.¹

It represents approximately 1 percent of all anorectal malignancies.^{5,6} Between 0.4% and 1.6% of all melanomas arise in the anorectal region, and the anal canal is the most frequent site of melanoma after the skin and retina.⁶ The malignancy occurs more frequently with advancing age, with peak incidence in the sixth and seventh decades. Most patients are female^{1,2}

The common initial symptoms are bleeding per rectum and/or pain, anal mass, pruritus, tenesmus or change in the bowel habits. The surgical management of patients with anorectal melanoma is controversial.⁷ Some authors have stated that wide local resection is the first choice for primary anorectal melanoma if negative margins can be achieved when this is technically feasible and complete tumor resection is impossible.^{2,4} Abdominoperineal resection should be reserved for large tumors where wide local excision is not technically possible.⁴ Radiotherapy and chemotherapy need to be further evaluated, and currently there is no standard chemotherapeutic regimen for anal malignant melanoma.

CASE REPORT :

We report a case of a 65 year old man who presented with a history of bleeding per rectum since 1 year, significant weight loss, alteration in bowel habits and some mass coming out of anal canal since 6 months which gradually increased in size.

Bleeding was associated mainly with defecation, and the blood was mostly bright red in colour with few episodes of dark clotted blood in between. Digital examination of rectum revealed a fleshy mass occupying almost whole of the anal lumen with protrusion of approximately 4 cm and reaching up to the anal verge. Sigmoidoscopy showed a ulcerative black colored nodular growth starting at anorectal junction, and extending proximally for 4 cm. The histopathological examination showed features suggestive of malignant melanoma, and was positive for HMB-45 and S100. CECT whole abdomen and pelvis revealed a fungating intraluminal mass in the rectum extending proximally just below the rectosigmoid junction to the anal verge distally with para-aortic and external iliac lymphadenopathy and metastasis to lungs. A diversion sigmoid loop colostomy was done to relieve obstruction, and was referred to an oncologist.



Figure 1. Patient with primary anal malignant melanoma before surgery

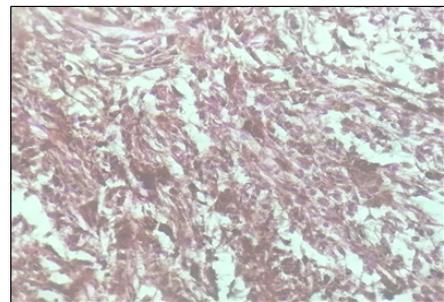


Figure 2. Histopathology showing clusters of malignant cells and abundant melanin pigment in cytoplasm

DISCUSSION:

Moore first described anorectal melanoma and its treatment with local excision in 1857. Anorectal melanoma is an uncommon and aggressive cancer with an unfavourable prognosis.¹⁻⁴ It is more common in females and the mean age of disease onset is 60 years.¹² It is characterized by unspecific symptoms, and differential diagnosis with other lesions of the rectum and anus is often difficult.

Malignant melanoma is a tumour arising from epidermal melanocytes originating from the neural crest. During fetal development, these cells migrate to many sites throughout the body, primarily to the skin. However, melanocytes also reside in the eyes (retina and uveal tract) and mucosal surfaces (head and neck, anorectum, female genitalia). Anal melanoma is the third most common melanoma after the cutaneous and ocular varieties. Lesions can affect the anal canal, the rectum or both, with the majority occurring within 6 cm of the anal verge.⁹ The melanocytes are subjected to carcinogenic stimuli and subsequently undergo malignant transformation into melanoma. It is well documented that the risk of cutaneous melanoma is increased with exposure to sunlight, particularly UVB radiation. As the anal mucosa is essentially never exposed to sunlight, it is not obvious what triggers the development of anal melanoma. Many appear without pigmentation, and 20% are truly amelanotic on histologic examination.^{7,10,11}

The common initial symptoms are bleeding per rectum and/or pain, anal mass, pruritus, tenesmus or change in the bowel habits. Macroscopically, the majority of these tumors are polypoid and pigmented and arise near the dentate line. They may also present as nodular prolapsed masses, as in our patient. Microscopically, the tumor cells are arranged in nests and individual cells may be epithelioid or spindle. These clusters of tumor cells invade the overlying squamous mucosa in a pagetoid manner and are characterized by immunostaining specific for the melanosome protein, HMB-45. If metastatic disease is present, the symptoms may include weight loss, anaemia, fatigue, groin masses, pelvic masses, or even bowel

obstruction. The symptoms of anal melanoma are also misdiagnosed as symptoms of other anorectal pathologies such as haemorrhoids, skin tags, or polyps.¹³ It is presumed that primary anorectal malignant melanoma arises from normal melanocytes in the intestinal epithelium distal to the dentate line and extending proximally onto the rectum.¹⁴

The staging of anal melanoma differs from that of cutaneous melanoma, which is based primarily on thickness in millimetres (Breslow classification). Anal melanoma is staged on a clinical basis, focusing on locoregional and distant spread. Stage I is local disease only, stage II is a local disease with regional lymph nodes, and stage III is distant metastatic disease.

The abundant lymphatics of the anorectum probably facilitate the high rate of inguinal and iliac lymph node metastases. The rich vascular network in this area promotes haematogenous spread to liver, lung, bone, brain, and other organs. Radiological investigations should be directed at these organs.

Malignant melanoma of anorectum is best evaluated by endoscopy and biopsy, although CT scan may prove to be a valuable tool in the assessment of regional diseases, especially if additional treatment modalities, such as chemotherapy or radiation therapy, are being considered. The use of immunohistochemistry panels, including S-100 proteins, MelanA, HMB-45 and tyrosinase can help in the diagnosis.¹⁵ The overall survival time is 10-19 months after diagnosis.^{2,7}

Surgical excision remains the primary treatment of anal melanoma. In addition, surgery plays an important role in palliative care and management of locally recurrent disease. The only uncertainty is the extent of excision, that is, a limited excision (wide local excision [WLE]) or radical excision (abdominal perineal resection [APR]). The surgical management of patients with anorectal melanoma is controversial.^{4,5,7} The prognosis of primary malignant anorectal melanoma is poor, irrespective of surgical treatment. A recent article has shown that the survival rate of patients with anal melanoma is similar after local excision or rectal resection, irrespective of whether patients have the localized or regional stage of the disease.¹⁶

Sentinel lymph node dissection may be indicated in clinical apparent disease or for occult primary. SLN dissection has been used in anorectal melanoma.¹⁷ A few case reports exist regarding the technical feasibility of SLN with anorectal melanoma,¹⁸ but the efficacy remains unknown.

Vaccines have been examined with some mixed results and not so sufficient data for anorectal melanoma.¹⁹

The role of chemotherapy for melanoma remains unclear. Biochemotherapy refers to the administration of both a biologic agent and a traditional cytotoxic chemotherapeutic agent. It represents the best available adjuvant therapy after surgery and should be considered in the treatment of advanced or metastatic anorectal melanoma.

The use of radiation as part of the management of anorectal melanoma has been reported. Adjuvant radiation therapy is well tolerated and is promising in improving locoregional control. Postoperative radiotherapy may improve locoregional control after wide local excision. Definitive assessment of the efficacy of adjuvant radiation therapy requires further prospective studies.²⁰

Five-year survival for all patients with anal melanoma is only 3 to 22%.²¹⁻²³ Survival in patients with recurrent or metastatic disease is less than 10 months.^{21,23} There are no reported cases of long-term survivors if metastatic disease, including inguinal lymph node metastasis, is present in this case due to presence of metastasis surgery was considered only for palliation and patient was referred to oncologist for further management.

CONCLUSIONS:

We should have differential diagnosis of malignant melanoma of anal canal in an old age patient presenting with a black coloured mass. Diagnosis should be confirmed by histopathology along with other investigation to evaluate the extent of the disease and such patients should be treated promptly either by wide local excision on abdominoperineal resection as these tumors are very aggressive in their course. Chemotherapy, radiotherapy, and immunotherapy should be considered in the treatment of anorectal melanoma in order to influence overall survival.

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