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Primary anal malignant melanoma: a case report

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ABSTRACT
Primary anal malignant melanoma is an uncommon and aggressive disease. In many cases, the disease is undetected or mistaken for a benign polyp or haemorrhoids until it reaches an advanced stage. 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 70 year old male who was managed with wide local excision of the growth

I. INTRODUCTION

Primary anal malignant melanoma is an uncommon and aggressive disease. Lesions can affect the anal canal, the rectum or both, with the majority occurring within 6 cm of the anal verge.1 Malignant melanoma is a tumour arising from epidermal melanocytes originating from the neural crest. Melanomas of the anorectum are the third most common after melanomas of the skin and retina. The malignancy occurs more frequently with advancing age, with peak incidence in the sixth and seventh decades and no apparent sexual predilection. The common initial symptoms are bleeding per rectum and/or pain, anal mass, pruritus, tenesmus or change in the bowel habits. Surgical excision is the primary treatment of anal melanoma including wide local excision, and abdominoperineal resection. Radiotherapy and chemotherapy need to be further evaluated, and currently there is no standard chemotherapeutic regimen for anal malignant melanoma.

II. CASE REPORT

A case of 70 year old male, presented to the out-patient department with a three months history of pain peri-anal region and bleeding per rectum, with some mass coming out of anal canal which gradually increased in size over 3 months. 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 firm polypoidal cauliflower growth of the size of 7 x 6 cm at the muco-cutaneous junction at 6'o clock position. There was no significant inguinal lymphadenopathy. USG abdomen showed rectal wall thickening. CT scan abdomen revealed a well-defined polypoidal, heterogeneous, moderately enhancing mass lesion seen at anal verge, also infiltrating anal canal distally, with few lymph nodes in para-aortic, mesenteric and peri-rectal region, and mild thickening of posterior rectal wall. Fat planes of anterior rectal wall was maintained. However there was no evidence of any distant metastasis. Patient's routine blood investigations were all within normal limits. The chest X-ray was normal. Stool for occult blood examination was negative. The patient was posted for surgery after confirmation of malignant melanoma in incision biopsy and wide local excision was done.

Upon excision biopsy of the growth, the resected specimen showed some pigmented lesions within the tumour and around the anal verge. The cut-section of the growth was brownish with some grey-white areas. The histopathological examination showed features suggestive of malignant melanoma.

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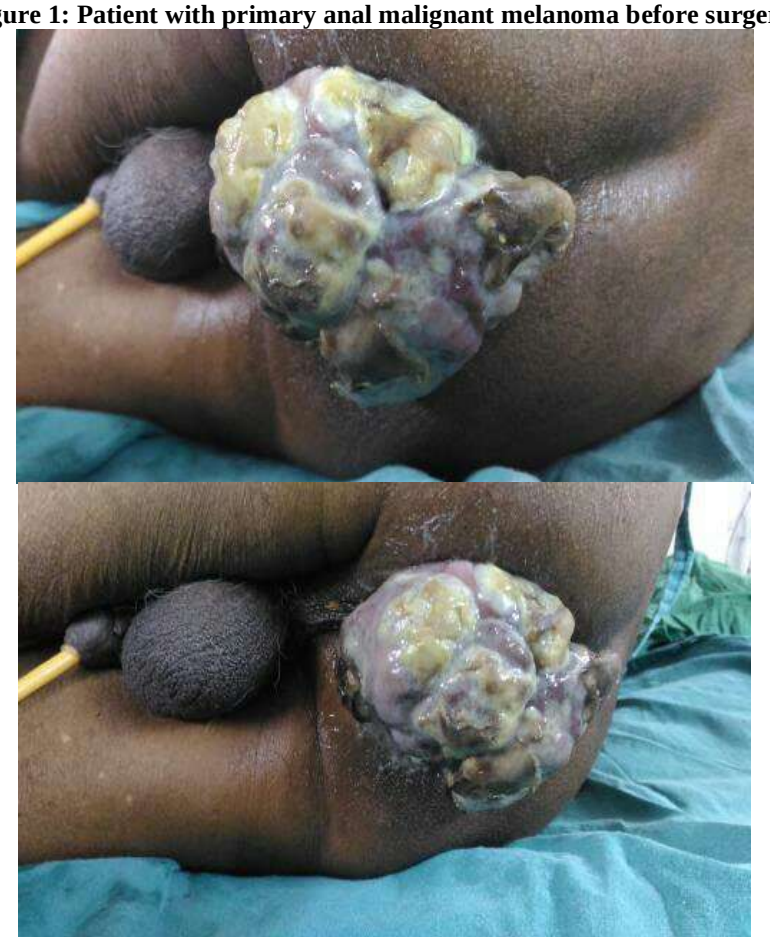
Figure 1: Patient with primary anal malignant melanoma before surgery.



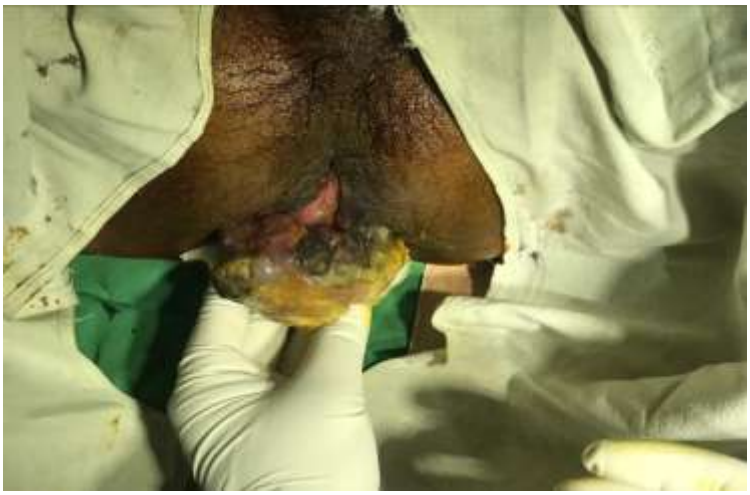


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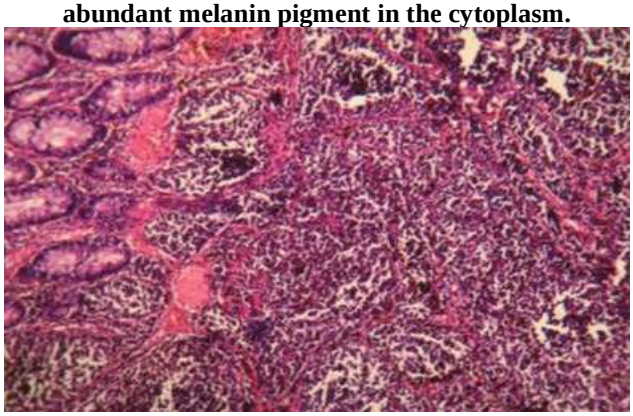
Figure 2: Patient with primary anal malignant melanoma after surgery.

Figure 3: Histopathology showing anal glands and clusters of malignant cells with pleomorphic nuclei and abundant melanin pigment in the cytoplasm.



III. DISCUSSION

A melanoma of anus and rectum was first reported by Moore in 1857.2 Lesions can affect the anal canal, the rectum or both, with the majority occurring within 6 cm of the anal verge .Malignant melanoma is a tumour arising from epidermal melanocytes originating from the neural crest. The melanocytes are the cell that synthesize melanin and are located in the basal layers. 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 the uveal tract) and mucosal surfaces (head and neck, anorectum, female genitalia). It is well documented that the risk of cutaneous melanoma increases with the exposure to UVB radiations. Malignant melanomas occur in the anorectum because of the presence of abundant melanocytes in the mucosa of the anal canal. As the anal mucosa is essentially never exposed to sunlight, it is not obvious what triggers the development of anal melanoma.

Malignant melanoma of the anorectum is rare and has very poor prognosis. The incidence has been reported to be 0.4%-3.0% of all malignant melanoma and 0.1%-4.6% of all anorectal malignant tumors.3-5 Melanomas of the anorectum are the third most common after melanomas of the skin and retina. The reported 5-year overall survival rate is 6%-15% of patients after surgery.3,6-11 Early-stage detection is important. The main determinants of prognosis are the depth of invasion and stage of the disease.

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 epitheloid or spindled. These clusters of tumor cellsinvade 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.12 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.13 The staging of anal melanoma differs from that ofcutaneous 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. In various series, 20% to 62% of patients had metastatic disease at the time of initial diagnosis. The abundant lymphatics of the anorectum probably facilitate the high rate of inguinal and iliac lymph node metastastes. The rich vascular network in this area promotes hematogenous 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.14 Surgical excision is the primary treatment of anal melanoma. In addition, surgery plays an important role in palliative care and management of locally recurrent disease. The majority of patients are diagnosed at a relatively late course in their disease, and therefore curative excision is simply not possible for these patients. The only uncertainty is the extent of excision, that is, a limited excision [Wide Local Excision (WLE)] or radical excision [Abdominal Perineal Excision (APR)]. Initial treatment suggested is WLE because radical surgery failed to show any survival advantage and also to avoid the need for colostomy.15,16 Local recurrences are common with WLE with no documented effect on survival. However, the rarity of this tumor, advanced stage at presentation, and poor prognosis have confounded attempts to clarify optimal surgical intervention.Most patients die regardless of the chosen therapeutic strategy due to the aggressive nature and the rapid progression of the tumour. 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 Chemotherapy role remains unclear. 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 No published randomized trials are available for anal melanoma due to the small number of patients requiring several decades to recruit in most series. Hence, all treatment regimens are extrapolated from trials involving metastatic cutaneous melanoma. Future options will likely incorporate combined modalities of chemotherapy, immunotherapy, and radiation therapy.

IV. CONCLUSION

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 alongwith other investigation to evaluate the extent of the disease and such patients should be treated promptly either by wide local excision or abdomino-perineal resection as these tumors are very aggressive in their course.

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