Anal Melanoma - a Case Report and Review of Literature

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Abstract

Malignant melanomas of the anal canal are very rare cancers with a very poor prognosis. These tumours behave very aggressively and have a negligible five-year survival, due to late diagnosis and early metastasis. Moore reported the first case in 1857. Melanomas account for less than 1% of all tumours of the anal canal. Anal melanomas represent 1% of all melanomas. Amelanotic melanomas of the anal canal have also been reported. 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. The role of sentinel lymph node biopsy for staging of anal melanoma remains unclear. Treatment options for anal melanomas include wide local excision or an abdominoperineal resection. The use of radiotherapy and biochemotherapy needs further evaluation and is the subject of ongoing trials that are few and far between, due to the rarity of the tumour. We report a case of anal melanoma that was treated by abdominoperineal resection.

INTRODUCTION

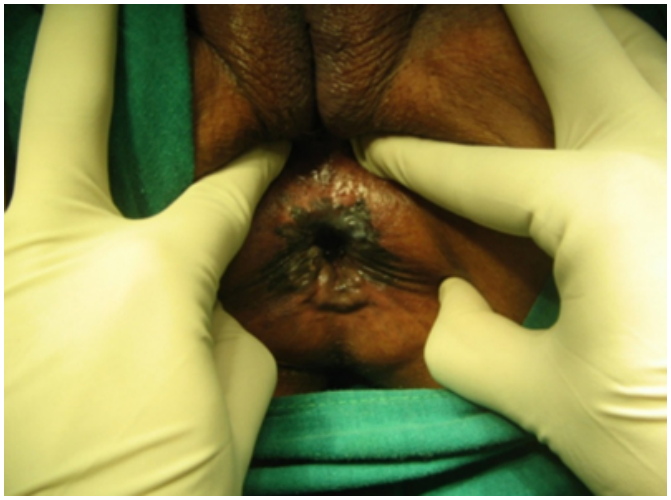
Anal melanomas are exceedingly rare tumours with a median five year survival of less than two years despite curative surgery[1] . At the Memorial Sloan Kettering Cancer Centre, 85 patients where seen in a 64 year period between 1929 to 1993. Survival was poor with a 17% five year survival (median, 19 months.) Besides occurring in the anal canal, melanomas most frequently originate from the mucous membranes lining the respiratory, digestive, and genitourinary tracts or in the eyes as well as in the cerebral meninges.[2] Females are more likely to be affected, generally presenting in the sixth or seventh decade.[3] Many of these patients masquerade as haemorrhoids, requiring therefore a high degree of suspicion to diagnose these tumours early. Unfortunately the tumour is widely metastasized at the time of initial diagnosis, making cure next to impossible. The Clark level and Breslow index used for evaluation of cutaneous melanomas are not applicable to extracutanous melanomas and, at present, there are no consistent, internationally accepted therapy standards for this form of the disease.[2] Surgical treatment modalities include wide local excision, and abdominoperineal resection. Radiotherapy and Chemotherapy need to be further evaluated, and currently there is no standard chemotherapeutic regimen for metastatic anal malignant melanoma.

CASE REPORT

We report a 65-year-old female patient who presented to the out-patient department with a six-month history of bleeding per rectum, and an extremely foul smelling blackish discharge in her undergarments. Abdominal examination revealed no abnormality besides a small para-umbilical hernia. There was no significant palpable inguinal lymphadenopathy. Per-rectal examination revealed a blackish discolouration of the peri-anal skin circumferentially around the anal orifice, extending for about a centimetre away from the anal orifice. There was no induration in this area; however, on doing a digital examination, induration was felt circumferentially all around the anal mucosa for a distance of two centimetres.

Figure 1

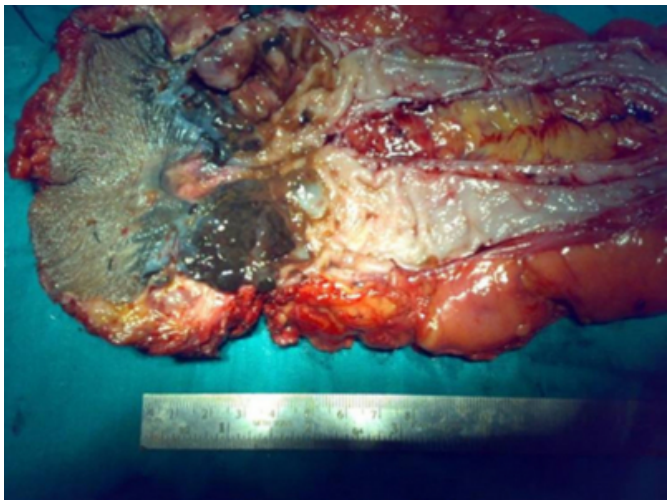
Figure 1: View of the anus showing perianal black pigmentation.



Proctoscopy revealed two polypoidal masses involving the anterior and lateral walls, and extending two centimetres proximal to the anal verge into the anal canal. There was also a blackish discharge with foul smell just as the patient had complained. The patient was admitted immediately. Routine blood tests were normal besides showing anaemia (Hb 9g%). Renal and liver function tests where normal. A punch biopsy was taken, which revealed a tumor showing malignant cells with pleomorphic nuclei and abundant melanin pigment in the cytoplasm reported as malignant melanoma of the anal canal. Colonoscopy did not reveal any other similar lesion or abnormality. An abdominal ultrasound and a contrast-enhanced CT scan of the abdomen were done. CT imaging showed a circumferential mass involving the anal canal for a length of 4cm. The growth showed endoluminal as well as exophytic components with breach of the serosa. The fat plane between the growth and lateral pelvic wall, uterus and bladder was maintained. There were no liver metastases, ascites or perirectal lymph nodes. The chest X-ray was normal. The patient was posted for surgery, and underwent an abdominoperineal resection (APR), with total mesorectal excision, with anatomical repair of the para-umbilical hernia under epidural and general anaesthesia.

Figure 2

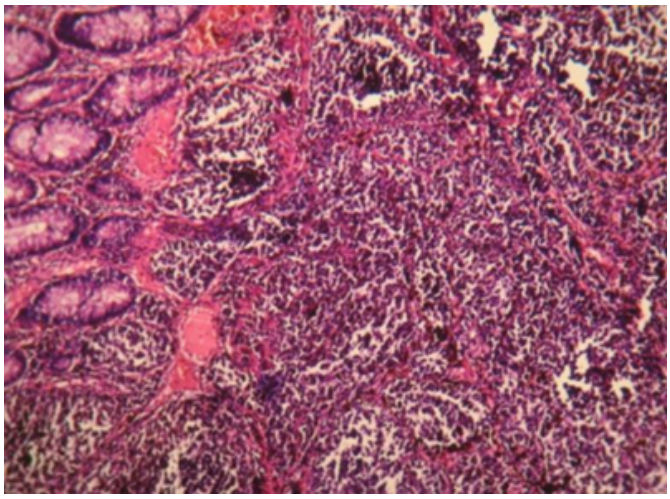
Figure 2: Specimen of APR showing the anal melanoma.



After a fairly stormy post-operative period, during which she developed a left basal pneumonia and a superficial abdominal wound infection, she was finally discharged home on the 21st post-operative day. Histopathology showed the same picture as the previous punch biopsy, with a satellite nodule measuring 1.5cm located 2cm away within the anal canal. Microscopically, the serosa and the periserosal fat were involved.

Figure 3

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



Three perirectal lymph nodes showed evidence of metastasis.

One month later, when she came for follow-up, all her wounds had healed, and there was no inguinal lymphadenopathy. She is presently undergoing work-up for

consideration of radiotherapy.

DISCUSSION

Malignant melanoma of the anal canal is an extremely rare and lethal tumor. The first case of anal melanoma was reported by Moore in 1857[4]. Prior to that, it had mainly been reported in horses. In normal individuals, melanocytes are readily identified within the squamous mucosa of the anal canal distal to the dentate line. Melanomas of the anorectum are thought to arise from these cells[5]. Macroscopically, the majority of these tumors are polypoid and pigmented and arise near the dentate line, as in our patient. They may also present as nodular prolapsed masses. Microscopically, the tumor cells are arranged in nests and individual cells may be epitheloid or spindled. 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-453[6].

Clinically, the patients present with bleeding per rectum or diarrhoea with tenesmus. Thus, malignant melanomas masquerade as haemorrhoids. The growth may ulcerate giving rise to severe pain on defecation. On examination, a tumour can be seen and can always be palpated, although on occasion the primary growth is small. Patchy perianal pigmentation may be noticed. Differential diagnoses include haemorroids, carcinoma of the rectum and prolapsing rectal polyp. There are times when the diagnosis appears straight forward, and at other times it is confusing thereby requiring a high degree of suspicion on the part of the treating physician. 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. In addition, anorectal melanomas often achieve large size and nodular growth before clinical detection. There is occult, widespread distribution of metastases before definitive evaluation of the symptomatic anorectal mass. Biopsy provides a means of absolute diagnosis.

While sentinel lymph node (SLN) biopsy remains the preferred method for nodal staging in cutaneous melanoma, its role for staging anal melanoma remains unclear.[7]

Despite refinements in surgical technique and emergence of various forms of adjuvant therapy, the overall prognosis of anal melanomas remains dismal. In spite of attempted curative surgery, the median survival for anorectal melanomas is only 20 months and most patients die within 5 years regardless of the type of intervention used.[8] From a review of literature, wide local excision (WLE) should be recommended where negative margins can be achieved and where this is technically feasible, thereby avoiding a permanent colostomy.[9] Endoluminal ultrasound can guide management by delineating lesions that are amenable to WLE.[10] Abdominoperineal resection (APR) should be reserved for large tumours where wide local excision is not technically possible. However, there is a school of thought that recommends APR in patients with localised anorectal melanoma, particularly in those with smaller tumours and no evidence of nodal metastases.[11] Historically, radical operations like APR with or without inguinal lymph node dissection were preferred. However, as no stage-specific survival advantage was seen with APR, later studies have recommended WLE as the preferred treatment where negative margins can be obtained.

In patients who have undergone resection with curative intent for anal melanoma, most recurrences occur systemically regardless of the initial surgical procedure.[12] 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.[13]

The combination of interferon (IFN), interleukin-2 (IL-2) and cytotoxic drugs, termed “biochemotherapy” or “chemoimmunotherapy,” seems to be more active than any agent alone, although no published randomised trials are available.[14] Biochemotherapy has been shown to have substantial activity against metastatic anorectal melanoma and should be considered for use in the treatment of metastatic disease from primary anorectal melanoma. Commonly used drugs include cisplatin, vinblastine, dacarbazine, interferon alpha-2b, and interleukin-2. All treatment regimens are extrapolated from trials involving metastatic cutaneous melanoma because of the very small number of patients of this very rare tumour that are available, requiring several decades to recruit in most series.

The use of vaccines to treat patients of anal melanoma, again obtained from data learnt from trials involving cutaneous melanoma, is based on the unique immunogenicity of this solid tumour. Preliminary studies have not shown any efficacy in the treatment of this dreaded tumour.[15] How very rightly it has been said that the sun never shines on anal

melanoma.

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