Malignant melanoma of the anal canal
A case report and review

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Summary
Primary malignant melanoma of the anal canal is a fairly uncommon but highly malignant disease. At the time of diagnosis it is usually far advanced and therefore incurable. Confusion with benign conditions and ignorance regarding the disease entity result in delayed diagnosis in a high percentage of cases. Improvement in the survival rate depends on acute surgical awareness, early diagnosis and aggressive surgical intervention.

Case report
A 79-year-old white man presented at Universitas Hospital, Bloemfontein, with an 11-day history of haematochezia, pain and tenesmus. Small amounts of bright red blood were passed during and after defaecation and sometimes as a blood clot in between. No appreciable amounts of mucus were observed but he had noticed that his motions were thinner. Pain was constant and of low intensity but was severely aggravated by passing stools, and tenesmus became an increasing nuisance. On two previous occasions, 15 and 8 months before, he had gone to his family practitioner with similar complaints. Each time a diagnosis of ‘severe piles’ was made and he was treated with suppositories and a blood transfusion.

This thin, elderly man appeared chronically ill. No abnormal masses, tumours or enlarged lymph nodes were found. Clinically he was anaemic with a haemoglobin value of 8.6 g/dl. An ECG showed ischaemic changes and unifocal ventricular ectopic beats. On rectal examination a raised, ulcerating and very painful tumour could be palpated. A sigmoidoscopy under general anaesthesia revealed a darkly pigmented, raised, ulcerating lesion which started about 3 cm from the anal verge; although clearly far advanced, no evidence of local or regional spread was noted.

After routine mechanical and antibiotic bowel preparation, an abdominoperineal (AP) resection was carried out by a combined approach. At operation the right internal iliac nodes were seen to be involved and were resected, but no other evidence of spread was found. The tumour was a darkly pigmented, raised, ulcerating lesion with a diameter of 8 cm (Fig. 1) extending from just distal to the linea dentata proximally into the rectum. On histological examination it was found to be a well-differentiated malignant melanoma extending through the bowel wall into the perirectal fat. Only internal iliac nodal involvement was seen.

On the 10th day postoperatively, after an uneventful recovery, the patient suffered a fatal coronary thrombosis. Autopsy showed no residual local tumour and no regional or distant spread.

Discussion
Primary malignant melanoma of the anal canal is a rare tumour, first reported by Moore in 1857. It comprises less than 1% of all malignant tumours in this region; after the skin and retina, the anal canal is the third most common primary site of melanoma. The ratio is about 1 melanoma for every 8 squamous cell carcinomas of the anus and every 250 adenocarcinomas of the rectum. It affects older people, the mean age of patients with this tumour being about 60 years. Very few have been reported in non-whites. Recent studies tend to...
show a female preponderance, as in other anal malignancies, \(^4,6\), although not all authors share this view. The symptoms resemble those of other anorectal malignant conditions, the most common being bleeding followed by pain, constipation, abdominal pain and body mass loss. \(^4,6\) Delay and difficulties in diagnosis frequently occur because the tumour is often mistaken for a benign condition such as haemorrhoids or anal fissure and because pigmentation is absent in 16 – 41% of cases. \(^3,5,6\)

It is generally accepted that anorectal melanomas originate in the melanocytes of the anal canal. \(^4,10,11\) In a detailed study of this region, Walls \(^11\) could not find melanocytes in the rectal mucosa, i.e. proximal to the linea dentata. There are cases that may qualify as primary rectal lesions, \(^12\) although the consensus is that they arise within the anal canal and, spreading upwards beneath the mucosa, may present as a rectal lesion. \(^6\) The tumour invades the submucosal plane of the rectum and spreads proximally without ulceration. The lesion seldom grows as much distally. \(^13\) The tumours are usually single, although Braastad \(^{et al.}\) \(^14\) found 14 out of 94 to be multiple. These 14 cases probably represent satellite nodules. The size can vary from a few millimetres to more than 10 cm and they can be polypoid, pedunculated, sessile or diffusely infiltrating. \(^7\) They have been described by Angeras \(^{et al.}\) \(^2\) to be nodular, acral lentiginous or superficial spreading on histological examination. Because of the small series no correlation between histological type and survival could be found. As might be expected, survival seems to be related to the stage of the tumour at the time of diagnosis. \(^7\) The advanced size of these tumours together with the absence of classic dermatological layers in this region makes Clark's classification useless. \(^6\) Because of the absence of a papillary dermis in this region Breslow's method seems to be the best way in which to evaluate these tumours. Equating thickness with distant spread seems reasonable since the only 5-year survivors found by Wanebo \(^{et al.}\) \(^5\) all had lesions less than 2 mm thick and were treated by an AP resection with or without a superficial inguinal node dissection.

Unfortunately metastases are common. Haematogenous metastases disseminate via the portal vein to the liver and in more advanced disease pulmonary, bone and brain metastases also occur. Efferent lymphatics from the anus and anal verge follow the lateral and downward rectal efferents to the internal iliac nodes. The rectum drains to the mesorectal nodes and then to the inferior mesenteric chain. Most of the lymphatic vessels from the anal cutaneous region terminate in the medial group of superficial inguinal nodes. Mesenteric nodal involvement is more common than that of inguinal nodes, \(^15\) and it may be that inguinal metastases occur only when lymphatic drainage dynamics are altered by tumour replacement of mesenteric nodes. Widespread satellitosis occurs in the scrotal and perineal skin with more advanced disease.

Surgical treatment has evolved from a conservative approach to the more aggressive one of AP resection, which gives a better overall long-term survival rate. Chiu et al. \(^6\) from the Mayo Clinic reported no survivors when radical surgical excision was not carried out. Prophylactic inguinal node dissection has been advocated.

At present it seems prudent to individualize management since the microstaging of the lesion provides the best correlation with survival. A radical surgical approach should be adopted for cure in patients with melanomas less than 3 mm thick. \(^5\) With more advanced lesions therapy should be aimed at local control of the disease. With a very large lesion, as in our case, AP resection may be the only way in which to effect local control. In view of the poor response to other treatment modalities, local recurrence should be treated surgically when possible.

Survival seems to be poor even with localized lesions. In the series reported by Chiu et al. \(^6\) there was a 11.8% 2-year survival, which seems to be the average at present. None of their patients had a recurrence after a 2-year disease-free interval, which suggests that a careful follow-up for at least 24 months is warranted. The only risk factors influencing prognosis are tumour thickness and evidence of nodal involvement at operation. At the present time the only hope for survival lies in early detection and an aggressive surgical protocol.

REFERENCES

1. Moore WD. Recurrent melanosis of the rectum after previous removal from the verge of the anus in a man aged sixty-five. Lancet 1887; ii: 290.