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Case Study

Anorectal Melanoma – A Report

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ABSTRACT

A review of the English language literature confirms that anorectal melanoma is a rare tumour with dismal prognosis. Delay in diagnosis and the inherent aggressive systemic activity of the tumour at the time of diagnosis have been postulated as factors contributing to the poor prognosis. Surgery has always been the mainstay of treatment with options of either a wide local excision (WLE) or abdomino-perineal resection (APR). Neither radiotherapy nor chemotherapy has shown encouraging results in terms of increasing survival or reducing loco-regional recurrence. Recently there has been much interest in the use of interferon-α following surgery. We hereby report two cases encountered during a period of 6 months and discuss the management of this rare condition together with a brief literature review.

Keywords: Anorectal melanoma, melanoma

INTRODUCTION

Anorectal melanoma was first described by Moore in 1857.[1] It is a rare tumour accounting for less than 1 % of all colorectal malignancies and less than 2 % of all melanomas.[2] Since the disease is uncommon, there has never been an accepted management protocol. Surgery, either in the form of a wide local excision (WLE) or abdomino-perineal resection (APR), has been advocated as the primary treatment. Neither has been shown to have any influence on survival and therefore the choice of surgery should be individualised. We report two patients who presented to us with anorectal melanoma and complement this with a brief review of the literature, pertaining in particular to the management of this condition.

CASE ONE

A 21-year-old Chinese male was referred to us from a private medical centre with a diagnosis of anorectal melanoma. The histological diagnosis was obtained from an incisional biopsy of a pigmented lesion in the anorectal region. His main complaint then was of a one-year history of intermittent fresh *per* rectal bleeding and a one-month history of alteration in his bowel habit.

He was clinically fit and examination of the abdomen was generally unremarkable. There was a pigmented lesion at the anal verge extending 1 cm into the perianal area. A mass was palpable on *per* rectal examination extending from the anal verge up to the dentate line. There were no enlarged inguinal lymph nodes.

His haematological and biochemical parameters were within normal limits and human immuno-deficiency virus (HIV) screening was negative. Computerised tomographic (CT) scan of the abdomen and pelvis revealed a 4 x 4 cm mass in the anal canal. The mass seemed to be very closely related anteriorly to the prostate. Two small para-rectal lymph nodes were also noted. The liver, spleen and pancreas were normal.

Colonoscopy performed confirmed that the pigmented lesion extended up to the dentate line. There was no other lesion found in the colon (*Fig. 1*). The diagnosis and treatment options were discussed and a surgical option in the form of an abdomino-resection (APR) was deemed suitable for this patient.

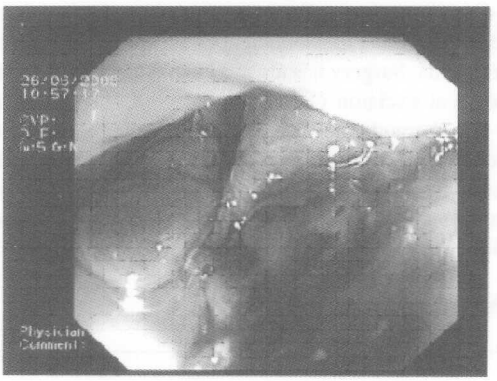

Figure 1. Pigmented mass in the anal canal on colonoscopy.

Intra-operatively, the tumour was found to be tethered to the prostate anteriorly. One pigmented para-rectal lymph node was noted. Post-operative recovery was uneventful. He was discharged well with a referral to the oncologist for adjuvant treatment.

CASE TWO

A 52-year old Chinese gentleman was admitted via the Accident & Emergency Department with worsening left iliac fossa pain within a period of three days. He claimed to have had episodes of intermittent rectal bleeding over the preceding three to four months associated with alteration in his bowel habit.

Clinically, he was noted to be pyrexial and in hypovolaemic shock. His abdomen was distended with a tender mass present in the left iliac fossa. Per rectal examination revealed a fleshy growth about 2 cm from the anal verge. A working diagnosis of a perforated large bowel tumour was made and he was aggressively resuscitated in view of impending surgery. Computer tomography (CT) scan of the abdomen and pelvis revealed a mass arising from the recto sigmoid region with a possible metastatic lesion in the left gluteal muscle.

An exploratory laparotomy was performed the next day following adequate resuscitation. Intraoperatively, there was an encapsulated mass arising from the recto sigmoid colon causing the large bowel obstruction. A sigmoid colesctomy along with resection of

the encapsulated mass was performed. Biopsies were taken from the anorectal mass noted preoperatively.

Postoperative recovery was unremarkable. The rectal biopsy was reported as primary malignant melanoma with the resected colonic specimen bearing a large peri-colic metastatic mass (Fig 2). The advanced stage and aggressive nature of the disease was discussed with the patient, and a decision was made for him to undergo palliative radiotherapy.

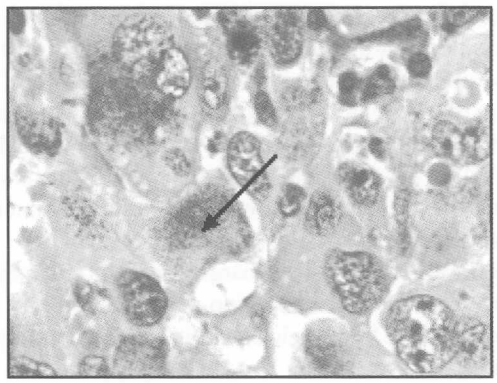

Figure 2. Pigmented melanoma cells as seen on the rectal biopsy

DISCUSSION

The overall incidence of anorectal melanoma continues to rise and a bimodal age distribution has been demonstrated[3]. Anorectal melanoma is commonly observed in the 25 to 44 age group and in patients older than 65 years. The recent high incidence of anorectal melanoma in the younger age group has been attributed to HIV infection. Human Immunodeficiency Virus infection has evidently been linked to development of other malignancies, even though the association between HIV and anorectal melanoma has never been conclusively proven. Nevertheless, it is presently recommended to screen all patients presenting with atypical anorectal malignancy for HIV infection.[3]

A series of patients with anorectal melanoma reported from Queensland, Australia, to determine the correlation between the high incidence of cutaneous melanoma to anorectal melanoma has failed to show any significant association. Anorectal melanoma was noted to be no more or less frequent in Queensland, despite a very high incidence of cutaneous melanoma. Unlike cutaneous melanoma, sun exposure is not a risk factor in the developmentof anorectal melanoma.[4]

The prognosis of anorectal melanoma remains dismal. In a retrospective series by Thibault et al, patients with anorectal melanoma treated primarily with surgery achieved a 5-year survival of only 22 % and a disease-free survival of 16 percent.[5] At the point of presentation, as high as 26 % of patients in their series, had metastatic disease. Delay in diagnosis was also considered to contribute to the poor prognosis. Most patients with

anorectal melanoma present with per rectal bleeding and this could wrongly be attributed to bleeding haemorrhoids. Due to this, there was significant delay from the time of presentation to the time of diagnosis. However, whether a timely diagnosis will have any impact on survival remains unproven.

Surgery either in the form of wide local excision (WLE) or abdomino-perineal resection (APR) has been accepted as the primary mode of treatment [5,6]. Historically, abdomino-perineal resection (APR) was thought to be the more appropriate surgical approach for potentially curable melanoma. Nevertheless, abdomino-perineal resection (APR) is a technically demanding procedure with considerable post-operative morbidity as well as leaving the patient with a permanent stoma. Wide local excision (WLE) is therefore an attractive option where patients can still be continent and avoid having a permanent stoma. Several authors have demonstrated that neither abdomino-perineal resection (APR) nor wide local excision (WLE) has any significant impact on both survival and locoregional recurrence. Patients who have undergone resection with curative intent developed systemic recurrence regardless of the choice of the operation [5,6].

Wide local excision (WLE) with 1-2 cm margins has been recommended as the operation of choice when technically feasible. Abdomino-perineal resection (APR) ideally should only be reserved for cases where free surgical margin is deemed difficult or for patients with isolated local recurrence.

We performed an APR for the first patient due to the anatomical location of the tumour, apart from the fact that the tumour was already invading locally into the perianal skin. The aim of the operation was curative and thus a sizeable margin had to be achieved. This proved to be appropriate since no local recurrence was noted twenty months after the surgery. The patient refused adjuvant treatment, including interferon therapy, and thus the effect of the treatment on distant metastases could not be ascertained. Multiple liver metastases were detected on the last computerised tomographic (CT) scan.

The second patient presented with an acute abdomen and the clinical features were very much suggestive of a neoplastic process. The main cause of the acute symptoms was basically due to the metastatic mass at the recto-sigmoid region rather than the primary mass in the rectum. This was resected but the primary mass in the rectum was only biopsied. Nevertheless this was considered adequate, bearing in mind the aggressive nature of this tumour, as well as the patient being in the older age group. Post operative radiotherapy only managed to control the bleeding but was not very successful in shrinking down the tumour. The patient finally succumbed to his illness ten months after the surgery.

Age, gender, extent of local infiltration, involvement of regional nodes and type of surgery have not been proven predictive factors of recurrence. It has been suggested that the only predictive factor for local recurrence is tumour thickness [6].

The role of radiotherapy, chemotherapy and immunotherapy in the treatment of anorectal melanoma remains controversial. Further evaluation in the form of prospective trials is recommended to address the issue. Recently there has been increasing interest in the use of interferon-α following excision of the tumour. Several reported series have demonstrated some advantages with this agent, even though a recent meta-analysis by Lens *et al.* demonstrated no clear benefit from using interferon-α [7]. Its role as adjuvant therapy in the management of anorectal melanoma should still be considered experimental.

CONCLUSION

Anorectal melanoma is an aggressive disease with poor prognosis. Regardless of the type of surgery, most patients succumb to the illness due to metastatic disease. The recent high incidence of anorectal melanoma in the younger population should alert the attending physician of the possibility of a concomitant HIV infection. Further studies are required to ascertain the role of new chemotherapeutic agents in the management of this potentially fatal condition.

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