

Primary Anorectal Malignant Melanoma: Clinical Features and Results of Surgical Therapy in Singapore—A Case Series

B S Ooi,**MBBS, FRCS (Edin), FRCS (Glas)*, K W Eu,***FAMS, FRCS (Edin), M Med (Surg)*, F Seow-Choen,***FAMS, MBBS, FRCS (Edin)*

Abstract

Introduction: Primary malignant melanoma arising from the anorectum is uncommon. The natural history of anorectal malignant melanoma is that of a very poor prognosis with early dissemination of disease. Successful surgical treatment has been rare. The present series reviews the clinical features and results of surgical management of patients with anorectal malignant melanoma treated in the Department of Colorectal Surgery, Singapore General Hospital. **Materials and Methods:** Data for all patients treated for anorectal malignant melanoma during an 11-year period from 1989 to 1999 were reviewed. The age, sex, presenting symptoms, duration of symptoms prior to diagnosis, size of tumour, extent of disease, type of surgery and length of survival were analysed. **Results:** Four men and 2 women, ranging in age from 31 to 81 years with histologically proven primary anorectal malignant melanoma, were included in the study. The most common (67%) presenting symptom was rectal bleeding. The mean tumour size was 2.5 cm (range 1 to 5 cm). All underwent abdomino-perineal resection. Three died of disseminated disease within 17 months while the other 3 were still alive at the time of this study; the longest up to 6.5 years from the time of diagnosis. **Conclusion:** The prognosis of primary anorectal malignant melanoma is poor. However, it is worthwhile treating aggressively as long-term survivor may be encountered in some.

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Key words: Abdomino-perineal resection, Malignant melanoma, Survival

Introduction

Malignant melanoma arising from the anal canal was first described in 1857. It is uncommon, comprising only about 1% of all anal canal tumours.¹ The anal canal, however, is the third most common site for primary malignant melanoma, superseded only by the skin and eyes.² It may be misdiagnosed as haemorrhoids or a polyp as it is often associated with non-specific symptoms such as rectal bleeding, anal pain or an anal lump.³ Up to one-third of anal canal melanoma are amelanotic or only lightly pigmented, making early diagnosis difficult.⁴ The natural history of malignant melanoma of the anal canal is that of a very poor prognosis with early dissemination of disease despite aggressive surgical and adjuvant therapy.⁵ To date, there is no universally proven effective adjuvant therapy for primary anorectal malignant melanoma. However, it is worthwhile treating aggressively as there may be some unexpected good results. Occasional long-term survivors following surgery with or without adjuvant therapy have been reported.³ Major series of anorectal malignant melanoma in the literature have come mainly from the Caucasian population. Furthermore, there is no report of

any long-term survivor in the Chinese population. All 4 Chinese patients with anorectal melanoma in a study from Hong Kong died within 5 years.⁶ We studied our experience with primary anorectal malignant melanoma.

Materials and Methods

Data for all patients admitted to the Department of Colorectal Surgery, Singapore General Hospital from April 1989 to December 1999 were prospectively entered into a computerised database. Information for primary anorectal malignant melanoma was extracted and analysed. The age, sex, presenting symptoms, duration of symptoms prior to diagnosis, size of tumour, extent of disease, type of surgery and length of survival were analysed (Table I). In all patients, the diagnosis had been confirmed by histology.

Results

There was a total of 6 patients (4 males, 2 females) with histologically proven primary anorectal malignant melanoma seen over an 11-year period (1989 to 1999). There were 4 Chinese, 1 Malay and 1 Bangladeshi. The mean age was 61.7 years (range 31 to 81 years). The most

* Associate Consultant

** Senior Consultant

*** Associate Professor, Head and Senior Consultant

Department of Colorectal Surgery

Singapore General Hospital

Address for Reprints: Dr Ooi Boon Swee, Department of Colorectal Surgery, Singapore General Hospital, 1 Hospital Drive, Singapore 169608.

TABLE I: ANORECTAL MELANOMA—CLINICAL DATA

Pt	Age (y)/ Sex	Presenting symptom	Duration of symptom	Size (cm)	Surgery	Stage	Recurrence/Metastases (mo)			Survival (mo)
							Local	Distant	Sites	
1	78/M	Bleeding	2 days	1	APR	II	Nil	10	Brain Lung Inguinal LN	12
2	67/F	Diarrhoea	1 month	5	APR	II	7	7	Pelvis Lung	8
3	31/M	Anal lump	1 week	2.5	APR	I	Nil	4	Inguinal LN	78*
4	81/F	Bleeding	1 month	1.5	APR	III	8	12	Lung Inguinal LN	17
5	71/M	Bleeding	3 days	2.5	APR	II	Nil	Nil	Nil	18*
6	42/M	Bleeding	2 months	2.5	LAPR	I	Nil	Nil	Nil	3*

APR: abdomino-perineal resection; LAPR: laparoscopic APR; LN: lymph node; Pt: patient; Stage I: localised disease; Stage II: nodal disease; Stage III: distant disease

* All 3 patients are still alive at the time of study

common presenting symptom was rectal bleeding (67%). Other symptoms were diarrhoea and presence of an anal lump. The diagnosis of anorectal malignant melanoma was suspected at initial presentation in all cases as they were all melanotic or pigmented. The mean duration of presenting symptoms prior to diagnosis was 20.7 days (range 2 to 56 days). The mean tumour size in diameter was 2.5 cm (range 1 to 5 cm).

Preoperative chest X-ray and computed tomography (CT) showed no evidence of metastatic disease in all patients except for 1 (patient 4) who had left inguinal lymph node metastasis, proven on fine needle aspiration biopsy. All patients underwent abdomino-perineal resection with wide excision of the perianal skin. Patient 4 had simultaneous bilateral groin block dissection while patient 3 underwent inguinal block dissection 4 months after the initial surgery.

The median length of survival was 12 months (range 3 to 78 months). Four (67%) of 6 patients had metastases to the perirectal lymph nodes (Stage II) at the time of surgery and 3 (75%) of these 4 patients died within 17 months. Patient 2 with the largest tumour (5 cm) had the shortest survival (8 months). Three patients were disease-free at a mean follow-up of 33 months (range 3 to 78 months). The youngest (31 years) was the longest survivor (78 months) despite initial left inguinal lymph nodes metastasis. There was, however, no correlation between the duration of symptoms or size of tumour with the length of survival (Table I). Local recurrence occurred less frequently (33%, 2/6) compared to the more common pattern of failures of

distant metastases (67%, 4 of 6).

Discussion

In our series of anorectal malignant melanoma, we found a preponderance of males (2:1). In contrast, all major series from Western countries have consistently reported a female preponderance (2:1).⁷⁻⁹ In addition, it has also been reported that up to 33% of anorectal malignant melanoma are amelanotic or only lightly pigmented.^{3,4} However, ours are exclusively melanotic lesions in contrast to reported literature. As our patients numbers are small, it is unknown whether there is any difference in the gender preponderance and degree of pigmentation for this tumour between the Oriental and Caucasian population.

Malignant melanoma of the anal canal is a rare and aggressive form of anorectal malignancy. It has a marked tendency for submucosal spread along the rectum with early metastases. A review of 267 patients with primary anorectal melanoma by Cooper et al¹⁰ showed that up to 62% of patients had metastases at the time of diagnosis. In our study, 67% of patients had metastases to the perirectal lymph nodes at the time of surgery. Perirectal, perianal and mesenteric lymph nodes are the most common sites of metastases, followed by inguinal lymph nodes, liver and lung. In a study of 50 patients over 55 years old at the Mayo Clinic,³ all patients who had metastatic disease at diagnosis died within 12 months (mean 6.3 months). In another study from St Mark's Hospital,¹¹ the mean survival was 8.8 months and only 2 (10%) of 21 patients survived beyond 18 months. In addition, among those patients who survived

more than 5 years after treatment, a majority of them developed local or distant metastases. The biological aggressiveness and delayed diagnosis probably accounted for the poor 5-year survival rate of 0% to 22% as reported in major series.³

However, there have been reports of surprisingly long survivors following surgery for primary anorectal melanoma. Quan et al¹² recorded 1 (5%) survivor out of 21 cases who was disease-free at 96 months who had a large 5-cm tumour at presentation, while Thibault et al³ reported 6 (12%) of 50 patients surviving beyond 5 years with a mean survival of 17 years (range 5 to 44 years). In our study, patient 3 survived and was disease-free for 78 months after an abdomino-perineal resection and bilateral inguinal lymphadenectomy.

Most authors^{3,10,11} found that survival was little influenced by the method of surgical treatment employed. There were no significant statistical differences in survival when patients treated by abdomino-perineal resection were compared with those treated by wide local excision.^{3,13} In general, radical surgery in the form of total rectal excision offers no more in terms of curative potential and little in terms of length of survival after palliative surgery. A less radical surgery such as wide local excision may, however, be more effective in control of local symptoms and postoperative continence. Therefore, wide local excision is recommended when technically feasible, because the prognosis is unchanged even by radical surgery.^{3,11,13} Abdomino-perineal resection is recommended if the tumour is too large, obstructing or not amenable to local excision.³ Abdomino-perineal resection for anorectal malignant melanoma performed laparoscopically, as for our patient 6, has not been assessed before. The role of prophylactic inguinal lymph nodes dissection is also unknown although Chiu et al⁷ found that all their 5-year survivors had undergone prophylactic inguinal lymph nodes dissection.

Gender, size of tumour, presence of melanin, positive perirectal lymph nodes or mode of treatment have all been shown not to be predictive of survival in patients with anorectal malignant melanoma.³ In a recent review of 117 cases of anorectal malignant melanoma, Cagir et al⁸ showed that the survival rate is better in young patients aged 25 to 44 years. However, young age has not been consistently shown to be a good prognostic factor by other studies. Staging by the TNM system has not correlated well with

prognosis as size alone does not correlate well with length of survival. Staging by the thickness of the original lesions, however, showed a better prognostic correlation than tumour size in some series.¹⁴ Quan et al¹² noted that no patient with a lesion of more than 1.6 mm thick survived 5 years. Wanebo et al⁹ observed that patients with lesions less than 2 mm in thickness survived, whereas those with lesions greater than 2 mm thick eventually died of their disease.

Our experience showed that primary anorectal malignant melanoma carries the same poor prognosis as in Western countries. The occasional long-term survivor may be encountered and all patients should be treated therefore.

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