# Malignant Melanoma of the Rectum

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## INTRODUCTION

Malignant melanoma of the anorectum is a rare and deadly condition. It accounts for 0.2%–3% (5) of patients with malignant melanoma, 0.1%–4.6% of anal malignant tumours of the rectum (9, 10). The prognosis is dismal, as fewer than 10% of patients survive more than five years despite operation (5–7).The anorectum is the most common site in the alimentary tract.The treatment not well defined as it is such a rare condition, the data must be collected over a long period, which makes statistical analysis difficult.

Surgical treatment of this virulent tumour ranges from the radical abdominoperineal resection with bilateral inguinal lymphadenectomy, to the conservative local excision alone. The optimal surgical treatment for individual patients is still controversial. Long-term survival is rare, as most patients die of disseminated systemic disease, regardless of treatment; for this reason some authors do not consider extensive radical surgery to be the treatment of choice. A recent study (7) found no difference in survival between patients who had had abdominoperineal resection and those who had had wide local excision. In our study we reviewed the published reports on the outcome of this disease, when treated surgically and the survival and recurrence rates after abdominoperineal resection and wide local excision to assess the relative merits of the two treatments.

## CASE REPORT

A 75-year-old woman presented with a history of recurrent rectal bleeding. During the previous year, the bleeding had become more frequent and severe. She also experienced episodes of alternate rectal bleeding and diarrhoea.

She had had cholecystectomy for biliary stones when she was 57.

Digital examination of the rectum showed a pedunculated polyp, 8 cm in diameter, in the anterior wall of the rectum. No lymph nodes were palpable. Laboratory data and tumour markers (CEA, CA19–9, CA195, and CA50) obtained at the time of admission were within the reference range. She was treated by wide local excision. The histopathological diagnosis was a muscle layer infiltrating melanoma, 1.4 cm deep, HMB45+ and S100+. The patient presented with distant metastases a year and six months after the operation.

## CONCLUSION

Melanoma is defined as "rectal" when it occurs in the rectum at an appreciable distance from the anorectal junction. In large series studies most patients had gross or histological pigmentation or both. Only 30% had amelanotic tumours. Large series studies (5, 8, 10), confirmed a female predominance of 2:1.

Rectal bleeding was the most common symptom at onset (2, 5, 7, 8). Sometimes the importance of the bleeding was underestimated as it was assumed to be a sign of haemorrhoidal disease, and this caused a delay in making the correct diagnosis and starting treatment.

Lesions are usually polypoid, only occasionally nodular or sessile, most being more than 2 cm in their largest dimension.

Because anorectal melanoma is often already advanced at the time it is diagnosed, earlier diagnosis is one approach to reducing mortality. One possibility is to look for melanoma, when screening for polyps or other common malignancies of the anorectum.

Regarding prognosis, in a review of 55 patients treated at the Mayo Clinic between 1973 and 1987 (10), the five-year survival was 22% and the disease-free survival was 16%.

Slingluff et al. (8) showed a five-year survival rate of 8%, but all survivors died within six years of the operation. Cooper et al. (4) reported that only 6.7% of patients who had treatment were free of disease five years after their operations. In the review from the Memorial Sloan-Kettering Cancer Center (3), 12% long term survivors and 9.4% long-term disease-free survivors were recorded. Some other series showed

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even lower rates of long-term survival and disease free survival (1, 4–6, 10).The factors that may account for the poor prognosis include: the advanced nature of the disease when diagnosed, ulceration, the rich vascularity of the rectal mucosa and heightened risk of haematogenous metastasis, and the probable high biological aggressiveness of these tumours.

High metastatic potential has been reported to explain the incidence of both diffuse visceral metastasis and massive pelvic extension.

Some authors have suggested that selection criteria such as stage of disease, thickness of lesions (less than 2 mm) or width of tumour free margin (greater than 2 cm), may be more important than the type of operation. A recent study (2) examined a few cases (18) immunohistologically using melanoma markers, epithelial markers, and DNA flow cytometry of paraffin-embedded blocks as well as other prognostic variables.

Conventional histopathological examination of paraffin-embedded specimens was made and tumour cells classified as: polyclonal, epithelial, spindle, small blue, and combinations of these. Ulceration, necrosis, blood vessel invasion, tumour thickness, and junctional or local components were also assessed.

For the immunohistochemical study the authors used the streptavidin-biotin-peroxidase method. They also considered polyclonal antiserum to carcinoembryonic antigen (CEA), broad-spectrum cytokeratin, low molecular weight cytokeratin, epithelial membrane antigen (EMA), vimentin, melanoma antigen, TAG-72, monoclonal antibodies to S-100, 3-amino-9-ethylcarbazole as chromogen (grading of staining: +3 = diffuse staining, over 50% of cells stained; +2 = intermediate staining, 10%–49% of cells stained; +1 = focal staining, 1%–9% cells stained; and + = rare, single-cell staining).

Flow cytometry was also done on paraffin-embedded specimens (DNA ploidy, S-phase, and $G_0/G_1$ peak) from which the authors concluded that aneuploidy of the tumour indicated highly malignant behaviour. Patients with thin tumours and an S-phase fraction of <10% had a trend towards longer survival. Nevertheless all these factors must be tested futher before they can be considered as reliable prognostic factors.

With regard to the treatment of choice, some authors (8, 9) found no difference in overall survival and the disease-free interval between patients who had curative abdominoperineal resection and those who were treated with wide local excision.

Notwithstanding those conclusions a study from the Memorial Sloan Kettering Cancer Center by Brady et al. suggested that abdominoperineal resection could be a reasonable approach for patients in whom the disease is not advanced, as this treatment was found to be more commonly associated with long-term survival than other procedures. They had not, however, made a statistical analysis of these data.

In conclusion, the low rates of survival given by the methods in use at present indicate that new approaches should be investigated. Adjuvant or neo adjuvant treatments might offer more effective ways of treating this disease. It is also important for clinicians to suspect anorectal melanoma so that earlier diagnosis may improve both the disease free interval and the overall survival of this uncommon but deadly disease.

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