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Anal melanoma: a rare, but catastrophic tumor

Andréa da Costa Veloso, Jaime Coelho Carlos Magno, José Antonio Dias da Cunha e Silva*

Colorectal Division of Hospital Naval Marcílio Dias, Rio de Janeiro, RJ, Brazil

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ABSTRACT

Introduction: Malignant melanoma of the anal canal is a rare and aggressive disease, which early diagnosis is difficult. Its presentation with no specific symptoms leads to a late diagnosis at an advanced stage. The prognosis of anorectal malignant melanoma is poor and frequently related to distant metastasis and absence of response of chemoradiotherapy. Surgery remains the mainstay of therapy; otherwise, the best approach is controversial. Considering no survival benefits for APR, wide local excision should be considered as the treatment of choice.

Methods: This report collects nine cases of anorectal melanoma treated at our division from 1977 to 2006, as well as a review of the literature.

Results: There were eight females and one male, of medium age 69 years (range: 41-85 years). Most frequent presentation was bleeding. Wide Local Excision (WLE) was performed in seven of them. Mean survival was 24 months, and six of them died on account of metastatic disease.

Conclusion: Anorectal melanoma remains challenging. Efforts should be taken to early diagnosis, and wide local excision with negative margins is the preferred treatment. Abdominoperineal resection (APR) is a reasonable option for bulky tumors or when the sphincter is invaded.

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Melanoma anal: tumor raro, mas catastrófico

RESUMO

Introdução: O melanoma maligno do canal anal é uma doença rara e agressiva, em que o diagnóstico precoce se torna difícil. Apresenta-se sem sintomas específicos, levando ao diagnóstico tardio e em fase avançada. O prognóstico é ruim e frequentemente relacionado a metástases a distância, bem como à ausência de resposta à rádio e à quimioterapia. A cirurgia permanece como terapia de escolha, no entanto a melhor abordagem ainda é controversa. Considerando não haver benefício na sobrevida da amputação abdômino-aerineal do aeto (AAPR), a excisão local ampla deve ser considerada o tratamento de escolha.

Métodos: São nove casos de melanoma anorretal tratados no serviço de coloproctologia do Hospital Naval Marcílio Dias (HNMD) de 1977 a 2006.

Palavras-chave:

Anal

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* Corresponding author.

E-mail: joseantoniocunha@yahoo.com.br (J.A.D.d. Cunha e Silva).

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Resultados: Foram diagnosticados oito mulheres e um homem, com média de idade de 69 anos (41 - 85). A queixa mais frequente foi o sangramento anal. A excisão local ampla foi realizada em sete pacientes. A sobrevida média foi de 24 meses.

Conclusão: O melanoma anorretal continua desafiante. Todos os esforços devem ser feitos para o diagnóstico precoce, tornando assim possível realizar a excisão local com margens negativas. A AAPR ainda é uma opção factível para tumores avançados ou quando o esfíncter anal está comprometido.

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Introduction

Anal malignant melanoma is a rare and aggressive disease with no specific clinical symptoms and a poor outcome. It is often diagnosed at an advanced stage, presenting with hematogenous metastasis. The anorectal region is the most common site for development of primary melanoma within the alimentary tract. The majority of the lesions arises from the dentate line of the anal canal and tends to spread submucosally.

Historically, anal melanoma was treated with abdominoperineal resection (APR), but recently some studies demonstrated no difference of the survival rates comparing to wide local excision (WLE). It appears that the only benefit of radical surgery is to obtain clear margins in tumor that are particularly bulky or invading the sphincter, and here APR should be considered. So, WLE offers the advantage of avoiding a permanent colostomy and is the preferred treatment when negative margins can be achieved.¹⁻⁴

Methods

The data of nine patients with anorectal melanoma were collected and retrospectively analyzed. From 1977 to 2006 we had a total of nine cases, including one male and eight females. Patients complaint, tumor characteristics, type of surgery, distant metastasis and overall survival were analyzed.

Due to the small number of patients, statistics analysis was not carried out.

Results

There were one male and eight females, of median age of 69 (41-85) years. Most frequent complaint was rectal bleeding. Two patients had amelanotic melanomas. Among nine patients, none presented with distant metastasis when the time of diagnostic. Six were Caucasian.

Seven patients underwent WLE, and one patient whose biopsy revealed melanoma refused any further treatment. This patient returned two years later with a bulky bleeding tumor, and a palliative sigmoidostomy was performed combined with embolization due to severe bleeding. One patient, at the time of diagnosis, presented as frozen pelvis, and had not enough time to a surgical approach. The median overall survival was 24 months. Among patients who underwent WLE we missed the follow up of one patient, one patient presented

a stroke and died five years after the procedure, and one patient is still alive (Figs. 1-6) (Table 1).

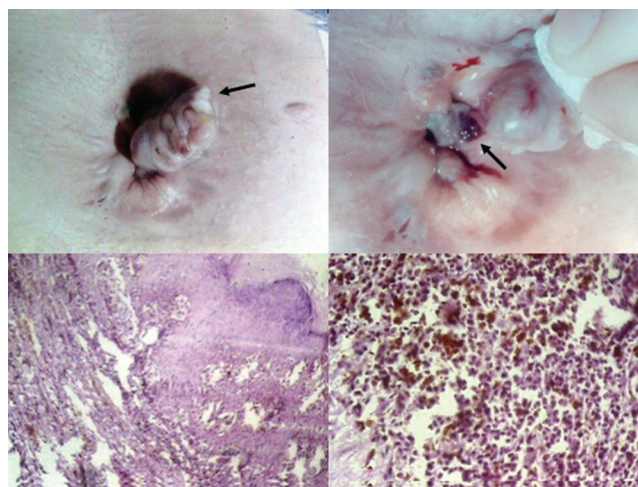


Fig. 1 – In clock hour: Nodular lesion with yellow pigmentation at the top (arrow). Dark pigmentation on the basis of the nodular tumoration. Intense cellular pleomorphism and melanotic pigment in the cytoplasm (case one).

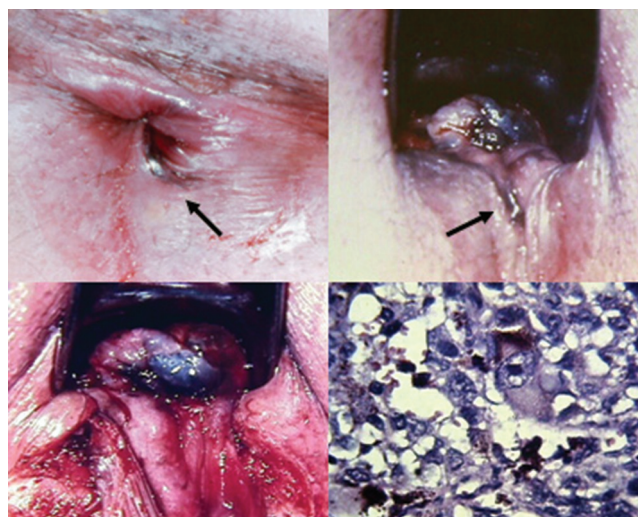


Fig. 2 – In clock hour: Dark pigmentation arising in anal verge extending to polypoid lesion in distal rectum (arrows). Wide local excision performed preserving the esfíncter muscle. Melanotic pigment in the cytoplasm of neoplastic cells (case two).

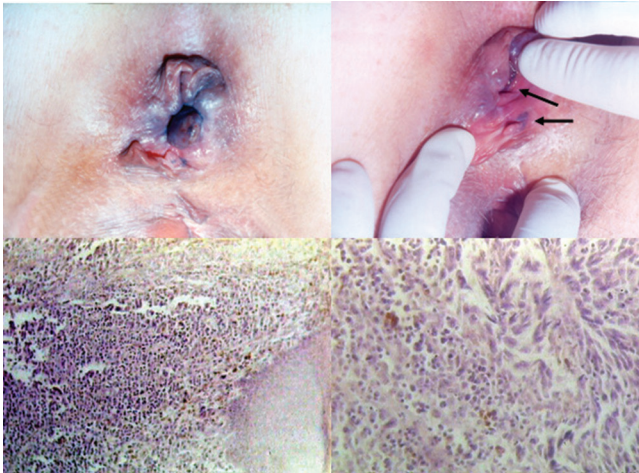


Fig. 3 – In clock hour: Typical dark nodular lesion in anal margin with black pigmentation (arrows). Histopathologic findings suitable of malignant melanoma (case four).

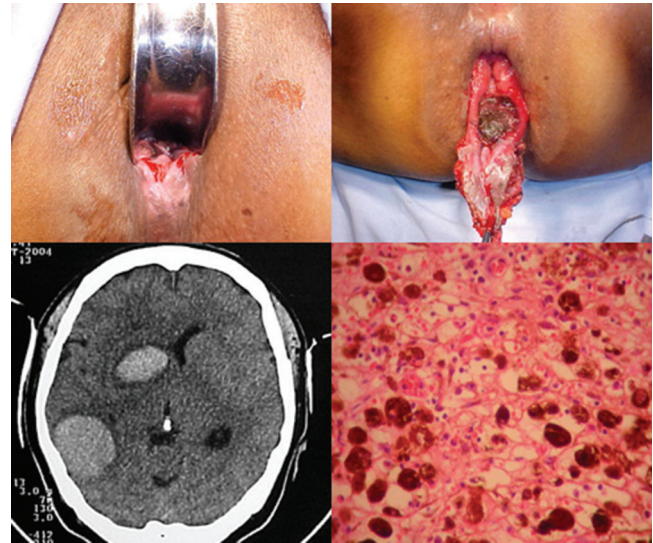


Fig. 5 – In clock hour: Fungiform dark mass in distal rectum. After neoadjuvant radiotherapy, wide local excision with loop ileostomy. Brain methastasis one month after surgery. Histopathologic findings show the lesion beginning at anal canal and extending to distal rectum (case seven).

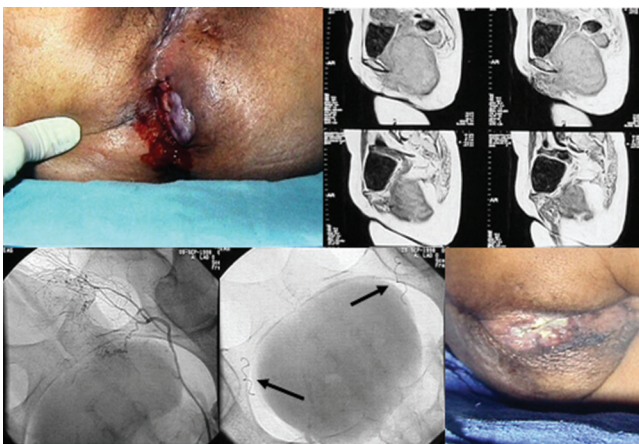


Fig. 4 – In clock hour: Vegetant and invasive lesion emerging from anal margin to pelvis, compromising rectal wall. Due to massive bleeding, it was performed arterial occlusion with coil (arrows). Late view after palliative radiotherapy (case six).

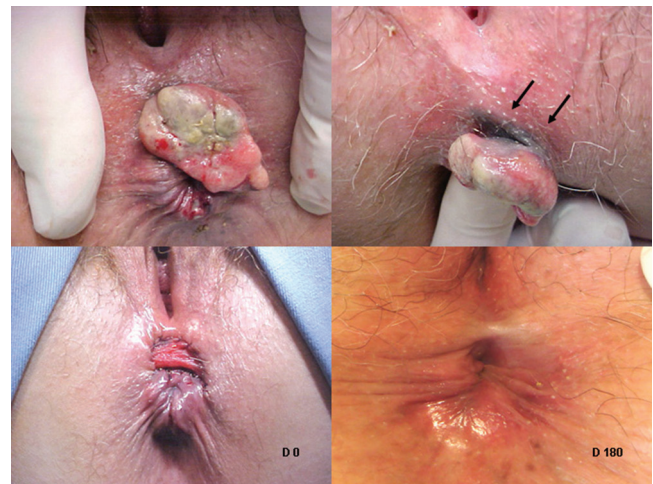


Fig. 6 – In clock hour: Nodular lesion with dark pigmentation at the basis in anal verge (arrows). Local excision with sphincter preservation and marsupialization of the surgical incision. Complete healing at 180 days (case eight).

Discussion

Anorectal malignant melanoma is a rare mucosal disease with a particularly aggressive biology compared with cutaneous melanoma of equal stage, accounting for approximately 1-4% of anorectal malignancies,^{2,5} but it is the third most common site after skin and eye and represents 0.6-1.6% of all melanomas.^{6,7}

The majority of patients are Caucasian, with the highest incidence during sixth and seventh decade. There is a slight female preponderance. The anorectal melanoma arises from melanocytes present in the transitional zone of the surgical anal canal and tends to spread submucosally.^{8,9} Related to its no specific symptoms, most of them, presenting with bleeding, prolapsed mass and anorectal pain, almost 60% of patients have already disseminated disease at initial diagnosis. Melano-

mas are commonly mistaken for hemorrhoidal disease, and the final diagnosis can be confirmed by positive excisional biopsy.^{1,2} The lesion is not pigmented in about 30% of cases and may resemble a villous carcinoma, and carries a worse prognosis by its invasive nature.^{2,7,9}

Immunohistochemical studies should always be done for establishing the diagnosis of melanoma. It is mostly positive for protein S-100 with a reported rate of 100%, melanoma antigen HMB-45 and vimentin.^{8,10} Others tests, as endoluminal ultrasound and computed tomography, should be performed to address the extension and presence of metastatic disease as its

Table 1 – Cases of anal melanoma.

| CASES | Year | Age | Gender | Local | Complaint | Tumor appearance | Surgery | Adjuvant therapy | Metastasis or recurrence (months) | Survival |
|-------|------|-----|--------|-----------|----------------|--|------------------------------|------------------|-----------------------------------|----------|
| 1 | 1977 | 71 | Female | Anal | Bleeding | Hemorrhoidal thrombosis | Local excision | Imuno + QT | Liver (24m) | 70m |
| 2 | 1977 | 69 | Male | Anorectal | Bleeding | Anal pigmented nodule | Local excision | QT | Rectum (6m) | 14m |
| 3 | 1993 | 58 | Female | Anorectal | Anal discharge | Rectal ulcerated mass | Local excision | QT + interferon | Lung (18m) | 23m |
| 4 | 1995 | 85 | Female | Anal | Bleeding | Anal pigmented nodule | Local excision | No | Groin (2m) | 29m |
| 5 | 1996 | 71 | Female | Anorectal | Bleeding | Rectal polyp | Local excision | No | Rectum lymphnode (8m) | 12m |
| 6 | 1996 | 42 | Female | Anorectal | Bleeding | Anal polyp | Sigmoidostomy + embolization | No | Liver (35m) | 40m |
| 7 | 2004 | 41 | Female | Anorectal | Bleeding | Rectal pigmented nodule | Local excision + ileostomy | RT | Brain (1m) | 8m |
| 8 | 2006 | 81 | Female | Anal | Nodule | Anal pigmented nodule | Local excision | No | No | alive |
| 9 | 2006 | 58 | Female | Anorectal | Bleeding | Rectal pigmented nodule and ulcerated mass | No | No | Frozen pelvis | 1m |

diagnosis. The computed tomography can reveals intraluminal masses in the distal rectum, with perirectal infiltration and often enlarged lymph nodes. Endorectal ultrasound is helpful in the evaluation of the tumor thickness and its nodal status.^{2,8}

Various factors, including duration of symptoms, inguinal lymph node involvement, tumor stage, the presence of amelanotic melanoma on histology, tumor necrosis, perineural invasion and tumor thickness, have been suggested to be negative prognostic factors. However, female sex has been suggested to be a positive prognostic factor for patients with cutaneous melanoma, and with an improved survival after radical resection for regional stage of anal melanoma. This reason is unclear.^{8,9,11}

The optimal treatment remains controversial, and surgery is based on two operative options: wide local excision (WLE) and abdominoperineal resection (APR). In the past, APR was advocated for the nonmetastatic disease, but as the prognosis was poor regardless of surgical approach, the goal of the surgical procedure should be based on obtaining negative margins and maintaining sphincter function. Most studies shows APR to provide a better local control, but no clear improvement in survival, and further consideration must be given to quality of life issues when making decisions between these two options.^{2,5,8,9,12,13,15}

Tumor thickness is a strong predictor factor for the risk of local recurrence and is used to plan therapeutic procedures. Tumor thickness below 1mm can be performed by local sphincter-saving excision with a 1cm safety margin. Tumor thickness between 1-4 cm should be excised by sphincter-saving excision and 2 cm safety margin, and tumor with thickness above 4 cm or with the involvement of sphincter should be treated with APR.^{8,9,11,13,14,18}

Stoidis et al. hypothesized that systemic dissemination is an early event in tumorigenesis, and by the time the lesion is clinically apparent micrometastases are well established. Metastases occur via lymphatic and hematogenous routes. Lymphatic spread to mesenteric nodes is more common than to inguinal nodes, while lungs, liver and bones are the most frequent sites of distant metastases.⁸

Prophylactic lymph node resection has no value, and the therapeutic lymph node resection should be performed only in the presence of positive inguinal nodes. Sentinel lymph node mapping (SLNM) has influenced the extension of surgical resection and seems to be helpful in preventing understating patients, who are pathologically node-positive but clinically node-negative and can allow early beneficial completion lymphadenectomy.⁵

The role of adjuvant therapy remains unknown. The response of anorectal melanoma to radio and/or chemotherapy continues to be poor. No systemic therapy regimen for metastatic anal melanoma is considered standard of care. Treatment is based on drugs developed for advanced cutaneous melanoma, although the clinical, biologic, and molecular seems to be different.⁸ Kim et al. used a combination of temozolomide, cisplatin and liposomal doxorubicin for metastatic anal melanoma with encouraging results.^{4,17}

Regardless of surgical approach, melanoma remains a highly lethal malignancy with overall 5-years survival rate less than 20%. The median survival is 34 months for patients with local disease and 10 months for those with metastatic disease.^{8,9}

The early diagnosis is the key to improved survival rate for patients with anal melanoma, due to the fact that the stage of the disease is the most important determinant in anorectal melanoma. A standard approach has not been established because of the limited number of patients of all anal melanoma reports. Further studies of the molecular mechanisms and tumor progression are needed to develop new treatment paradigms and improve survival.

Conclusion

Considering that anal melanoma is an aggressive tumor often diagnosed in advanced stages, a wide local excision should be considered as a therapeutic approach, despite controversial.

There's no significant impact in general survival with APR. So, WLE has its benefits in quality of life.^{13,16}

Conflicts of interest

The authors declare no conflicts of interest.

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