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Melanoma of the Anus Disguised as Hemorrhoids: Surgical Management Illustrated by a Case Report

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Introduction

Anal mucosal melanoma is a rare tumor that constitutes only 0.4–1.6% of all melanoma manifestations [1]. Prognosis is very poor, with a median survival of less than 2 years, despite curative surgery [2, 3]. Females are more likely to be affected than males and most patients present in the sixth or seventh decade of their lives [1]. Diagnosis is not always straightforward, and is often accidental after surgical treatment for presumed benign disease such as hemorrhoidectomy or lateral internal sphincterotomy, provided that tissue is submitted to the department of pathology [4]. Unfortunately, the tumor is often widely metastasized at the time of initial diagnosis [5, 6]. Here we present a patient who was referred to the surgeon for treatment of hemorrhoids but was diagnosed with anal melanoma.

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Case report

A 62-year-old black man with a history of prostatism was referred to our hospital by his general practitioner with the diagnosis of bleeding hemorrhoids. For a 10-week period he noticed a swelling protruding from the anus with daily bleeding from the anus. Defecation was problematical because of pain, but the stool had a normal aspect. Inspection of the anus showed a painful swelling with a diameter of 1.5 cm with a dark necrotic aspect surrounding the anus below the dentate line. Rectal palpation was impossible because of pain. At the outpatient department the swelling was excised under local anesthesia of the entire anus (anal block). Because of the atypical aspect of the swelling, the material was submitted for pathologic examination, which revealed a melanoma of the anus. Further staging of the tumor was not possible based on this material because of fragmentation of the tissue.

A sigmoidoscopy was performed but showed no further lesions. In addition, an abdominal and thoracic computed tomographic (CT) scan with contrast showed no signs of tumor around the anorectal region and no signs of intraabdominal, lymphatic or thoracic metastases. After the patient was diagnosed with having melanoma of the anus, examination under general anesthesia was performed. Inspection of the anal mucosa showed two additional lesions at 2 and 3 o'clock with the sacrum at 6 o'clock (Fig. 1). A wide local excision (WLE) including these lesions was performed with a margin of 2 cm, en bloc with 25% of the circumference of the rectal wall (Fig. 1). The total excised tissue was 6×7 cm. Pathologic examination revealed two small foci of the melanoma with a depth of maximal 3.5 mm. All margins were free of tumor. The patient recovered well from the operation. After 1-year follow-up by means of outpatient department visits and proctoscopy every 3 months, and thoracic and abdominal CT-scan twice a year, there were no signs of recurrent tumor.

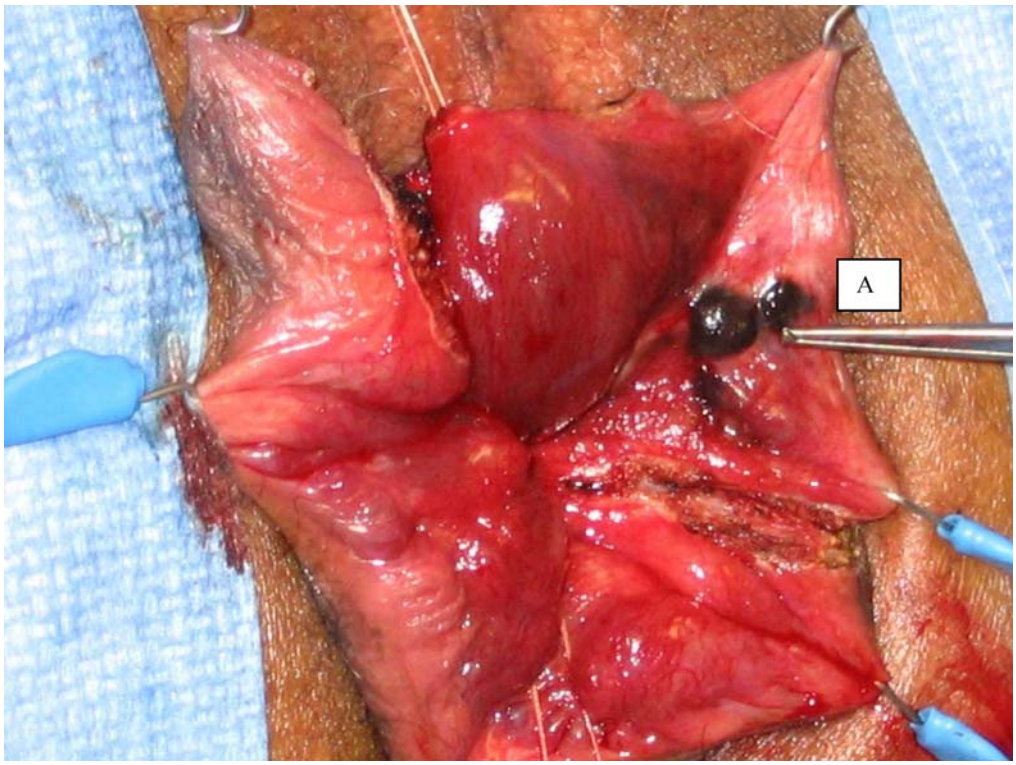

Fig. 1 Melanoma of the anus: wide local excision. A = melanoma

Discussion

Patients with anorectal melanoma usually present with symptoms of rectal bleeding and an anorectal mass and are often misdiagnosed as having hemorrhoids, as was illustrated by the present case report [4]. Therefore, on any atypical anorectal lesion, biopsy should be performed in order to prevent delayed diagnosis.

For the operative management of anorectal melanoma, two options are available: a wide local excision (WLE) or a more extensive abdominal perineal resection (APR). The choice between these surgical procedures is controversial. The main arguments in favor of APR are its ability to control lymphatic spread and to create bigger excision margins, resulting in an assumed lower local recurrence rate. But, in contrast to WLE, APR is associated with mortality, considerable postoperative morbidity (4% hemorrhage, 11–16% wound infection and 14–24% wound dehiscence) and the need for a permanent colostomy [7–11].

Despite attempted curative surgery, the median survival for anorectal melanoma is only 20 months and most patients die within 5 years regardless of the type of intervention used [12]. Therefore, quality-of-life issues must be given consideration when making treatment decisions.

Droesch *et al* performed a systematic review of the literature, including 14 studies with a total of 301 patients [12]. Wide local excision was performed in 129 patients, and 172 patients underwent APR. Local recurrence developed in 47% of patients after WLE and in 23% of patients after APR. Although these data suggest a trend towards better locoregional control after APR, the difference was not statistically significant. In addition, there was no difference in overall survival between WLE and APR (median survival 21 months for WLE and 17 months for APR).

Bullard *et al* found an opposite trend with a rate of 50% of local recurrence after APR, compared to 18% after WLE in 15 patients [5]. Postoperative radiotherapy may improve locoregional control after wide local excision. In the retrospective study of Ballo *et al*, actuarial 5-year local control was 74% and nodal control was 87% in 23 patients who received postoperative radiotherapy after wide local excision [13]. Definitive assessment of the efficacy of adjuvant radiation therapy requires further prospective studies.

In general, it is assumed that the stage of the disease, especially the tumor thickness, is the main prognostic factor [13–15]. Therefore, Weylandt *et al* suggested that the decision between WLE and APR should be governed by the tumor thickness [16]. In patients with a tumor thickness below 1 mm, a local sphincter-saving excision with 1 cm safety margin would be appropriate; and in cases of a tumor between 1 and 4 mm, a wide local excision with a margin of 2 cm seems to be adequate. Patients with a tumor thickness above 4 mm or invasion of the internal sphincter muscle should be treated with APR. In the case report mentioned

in this article, the tumor depth was 3.5 mm and excision margins were free of tumor, thus no additional APR was performed.

In order to evaluate the deepness of the lesion preoperatively, endorectal ultrasonography can be performed [6]. Accuracy in evaluating tumor depth of rectal cancer ranges from 81 to 94%, and accuracy in detecting lymph node metastasis ranges from 58 to 80% [17]. However, its accuracy in evaluation of anorectal melanoma remains unproved and is still evaluated [5].

At the time of diagnosis, up to one-third (16–33%) of the patients have disseminated disease [3]. For these patients, palliative treatment with chemotherapy might be a treatment option. Kim *et al* reported a series of 18 patients with metastatic anorectal melanoma treated with cisplatin, vinblastine, dadabazine, interferon alpha-2b and interleukin-2 [18]. Major response was seen in 44% of the patients, and complete response occurred in 11%, with a median overall survival of 12.9 months. Yeh *et al* used a different treatment regimen of temozolomide, cisplatin and liposomal doxorubicin in a patient with stage IV anal mucosal melanoma [19]. This patient showed a remarkable response to chemotherapy, with minimal residual disease and excellent quality of life at 12 months after the start of treatment. However, this therapy still has to be further evaluated and currently no standard systemic therapeutic regimen exists for metastatic anorectal melanoma.

In conclusion, anorectal melanoma represents both a diagnostic and therapeutic challenge to physicians given its non-specific presentation and rarity. It is associated with poor prognosis, regardless of the type of intervention used. Therefore, the overall treatment goal should be to optimize the quality of life. Since wide local excision is a more limited intervention associated with at least comparable survival compared to APR and no need for permanent colostomy, wide local excision is recommended as primary therapy if negative surgical margins can be achieved. APR should be reserved for patients in whom the tumor is thicker than 4 mm and/or involves the anal sphincter.

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