

Primary Anal Malignant Melanoma

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Abstract

Primary melanoma of the anal canal is a rare pathological event and comprises approximately one percent of all invasive tumors in this site. This devastating malignancy is often mistaken for benign conditions as either hemorrhoids or rectal polyp. Unfamiliarity with this bleeding anorectal lesion can lead to delay in diagnosis and therapy. As with melanomas elsewhere, the most useful characteristics for differentiating anorectal melanoma from other anal malignancies are melanin production, a "nesting" growth pattern and junctional changes. The prognosis is poor with a 5-year survival rate of about 15% inspite of an aggressive, multimodality approach. We present such a case with a detailed review of the literature.

INTRODUCTION

Primary anal malignant melanoma is a fairly uncommon but highly malignant disease. This disease is often mistaken for benign conditions, i.e., hemorrhoids or rectal polyp as they are the most common cause of anorectal bleeding₁. The epithelial lining of the anal canal is of adenomatous type in the upper part and is squamous in the lower part. The middle zone, also known as the anal transitional zone (ATZ), is characterized by an epithelium which bears resemblance to that of the anal glands, but show little mucus secretion₂. The melanocytes as demonstrated recently by melanocyte specific antibody (HMB-45) are usually located in the anal squamous zone and not in the colorectal zone, as was thought before. But in tumorous conditions, there is demonstration of melanocytes in all the three zones, which is interpreted as a tumor-induced proliferation of benign melanocytes, which are normally present, but in very small numbers or 'masked'. Thus the demonstration of melanocytes in all three zones of the anal canal substantially supports the observation that malignant melanoma of the anal canal may originate not only below, but also above the dentate line₃.

CASE REPORT

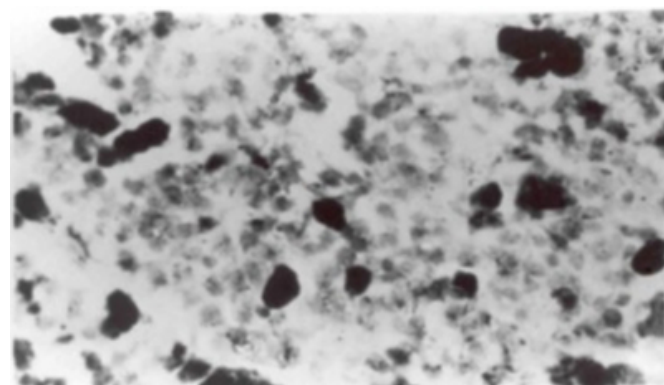
A 62-year-old, emaciated male presented with a history of bleeding per- rectum and gradually increasing difficulty in defecation of a six months' duration.

On examination, his general condition was poor. Pallor was present but there was no supraclavicular or inguinal lymphadenopathy. An abdominal examination was normal.

Per-rectally, there was a round globular mass arising from the posterior wall of the anal canal, about 2 cm from the anal verge which was hard, mobile, intraluminal and was not actively bleeding. A punch biopsy was done, which revealed a tumor showing malignant cells with pleomorphic nuclei and abundant melanin pigment in the cytoplasm (Fig. 1). Hence a diagnosis of malignant anal melanoma was made.

Figure 1

Figure 1: Photomicrograph showing malignant cells with pleomorphic nuclei and abundant melanin pigment in the cytoplasm (H&E, x 400).



Hematological investigations showed anemia (Hb=8g/dl). Other parameters [Liver Function Tests (LFT), ELISA for HIV, Australia antigen (HBsAg)] were normal. Ultrasonography (USG) of the abdomen was not contributory. Computed tomography (CT) scan of the abdomen and pelvis revealed a growth in the posterior anorectal wall, well preserved retroanorectal fat planes, but no local spread to adjacent organs. There was no mesenteric

lymphadenopathy or ascites. A colonoscopy showed a polypoid, pigmented lesion adjacent to the dentate line and the rest of the large bowel was normal. Considering these findings, the growth was deemed operable and the patient was subjected to an exploratory laparotomy, which confirmed all the above findings. An abdominoperineal resection (APR) was carried out. The resected mass was about 9 x 6 cms and the cut specimen revealed a blackish tumor mass (Fig. 2).

Figure 2

Figure 2: Gross resected specimen showing black pigmented tumor.



The final histopathology report was similar to that of the punch biopsy and the growth was not infiltrating the muscularis mucosa with no regional (mesenteric) lymph nodal metastases. No adjuvant therapy was planned due to the early stage of the disease. The patient has been kept under close follow-up since 2.5 years and is doing well with no symptoms or signs of distant metastases.

DISCUSSION

Primary anorectal melanoma comprises 0.25% to 1.25% of all the malignancies originating in this anatomic region⁴. Of all melanomas, anal melanoma represents 0.4% to 1.6% and is the third most common site of origin, following the skin and eye. It is usually a disease of old age, affecting mostly the fifth to seventh decade of life. There is no sex predilection but cases in whites outnumber blacks⁵. A recent analysis of 117 cases spanning 20 years in the National Cancer Institute registry showed a sharp rise in incidence in young men in San Francisco between 1988 and 1992, suggesting a possible association with HIV infection⁶.

Macroscopically, the majority of these tumors are polypoid and pigmented and arise near the dentate line, as in our patient. It may also present as a nodular prolapsed mass. Microscopically, the tumor cells are arranged in nests and individual cells may be epithelioid or spindled. These clusters of tumor cells invade the overlying squamous mucosa in a pagetoid manner and are characterized by immunostaining specific for the melanosome protein, HMB-453.

Surgical therapy in the management of anal melanoma is uncertain and controversial, but depends on the clinical stage of the disease. Procedures include a conservative approach of wide local excision (LE) and a more radical approach of APR. However, the rarity of this tumor, advanced stage at presentation and poor prognosis have confounded attempts to clarify optimal surgical intervention. Local lesions (without palpable lymph nodes) are treated by APR with or without groin dissection, whereas in an advanced disease with distant metastases and large primary, palliative treatment (local segmental resection, colostomy in intestinal obstruction) along with chemo-radiation may be done⁷.

It is noteworthy that no statistically significant difference has been demonstrated for APR (with or without inguinal node dissection) compared with wide LE. Poor prognosis was evident in each series, with a 5-year survival of less than 20%, regardless of the operative method. Surgical decisions must therefore be individualized according to the severity of the local symptoms, prevalence of distant metastases and overall well-being of the patient. Recent reviews advocate sphincter sparing LE, because patients tend to succumb to metastases regardless of surgical therapy^{4, 5, 6, 8}. In recent series, 20% to 62% of patients had metastatic disease at the time of initial diagnosis. The abundant lymphatics of the anorectum probably facilitate the high rate of inguinal and iliac lymph nodal metastases. The rich vascular network in

this area promotes hematogenous spread to the liver, lung, bone, brain and other organs^{5,9,10}.

A limited number of chemotherapeutic agents have shown effectiveness in metastatic cutaneous melanoma¹¹. No published randomized trials are available so far for anal melanoma due to the small number of patients requiring several decades to recruit in most series. Hence all treatment regimens are extrapolated from trials involving metastatic cutaneous melanoma. Future options will likely incorporate combined modalities of chemotherapy, immunotherapy and radiation therapy. The use of vaccines to treat patients of melanoma is based on the unique immunogenicity of this solid tumor¹². Preliminary studies have not shown any efficacy in the treatment of this tumor.

Despite the dismal prognosis of melanomas, there are a few isolated reports of lengthy survival¹³. The factors predictive of prolonged survival in these unusual cases are unknown but may be correlated with the depth of tumor invasion or with the absence of distant metastases at the time of diagnosis^{9,10}. As anorectal malignant melanomas are frequently advanced at the time of diagnosis, early diagnosis is critical to reduce the mortality rate.

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