

Primary Anorectal Melanoma: A Rare Case Report with Review of Literature

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Abstract

Anorectal melanoma is very rare malignant tumor, accounting 0.2-1% of all malignant melanomas and has a poor prognosis. The median survival of 8-23 months after the time of diagnosis. The typical treatment modalities includes surgery radiotherapy and chemotherapy. We present the case of a 75 year-old female with history of bleeding per rectum for 5 months with rectosigmoidoscopy showing an exophytic growth measuring about 5cm in diameter, multiple biopsies were taken showing tiny foci of submucosal infiltration by malignant cells. Abdomino-perineal resection was performed and pathologic examination revealed anorectal melanoma. Early diagnosis and treatment of this aggressive tumor for better out come and long term survival is highly recommended.

Keywords: Primary anorectal melanoma, abdomino-perineal resection.

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Introduction

Primary malignant melanoma of the anus and rectum is a rare and very aggressive neoplasm, constituting less than 1% of all melanomas and 4% of anorectal tumors other than adenocarcinoma^(1,2). Owing to its rarity and histologic variability misdiagnosis as carcinoma, sarcoma and lymphoma is common^(1,14). Sometimes this can be mistaken for benign conditions like hemorrhoids or rectal polyp as they present with rectal bleeding. The melanocytes as demonstrated by HMB-45 are usually located in the anal squamous zone, but are also seen in transitional and colorectal zone. Thus the demonstration of melanocytes in all the 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 line^(3,12). We present this case of this rare tumor in this location also we reviewed the literature related to the condition.

Case Report

A 75-years old female presented with 5 months history of bleeding per rectum. She was a known case of hypertension. By digital rectal examination, there was a mass about 2 cm from anal verge at five o'clock. Recto-sigmoidoscopy showed an exophytic growth measuring 5cm in diameter. Biopsy showed submucosal infiltration by malignant cells. Abdominal ultrasonography, computed tomography(CT) thorax showed no evidence

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of distant metastasis. An abdominoperineal resection was performed with permanent colostomy.

Grossly specimen received fixed in formalin composed of anus, rectum and sigmoid with a polypoidal shaped mass measuring 5x5x1.5 cm, away from the anus by 1.5 cm (**Figure 1**). Multiple lymph nodes were retrieved. Microscopic examination revealed malignant tumor arising and infiltrating the rectal and anal mucosa with deep infiltration through the muscularis propria. The tumor composed of clusters and sheets of cells with

round to oval nuclei and prominent eosinophilic nucleoli with melanin pigmentation of the cytoplasm and showed many mitotic figures (**Figure 2-a,b**). Immunohistochemical study was done and revealed positivity for melan A (**Figure 3-a**), S100 (**Figure 3-b**), and vimentin. While the tumor cells were negative for LCA and pankeratin, confirming the diagnosis of malignant melanoma. Two out of seven identified lymph nodes showed metastatic deposit.



Figure 1. macroscopic image of rectal melanoma

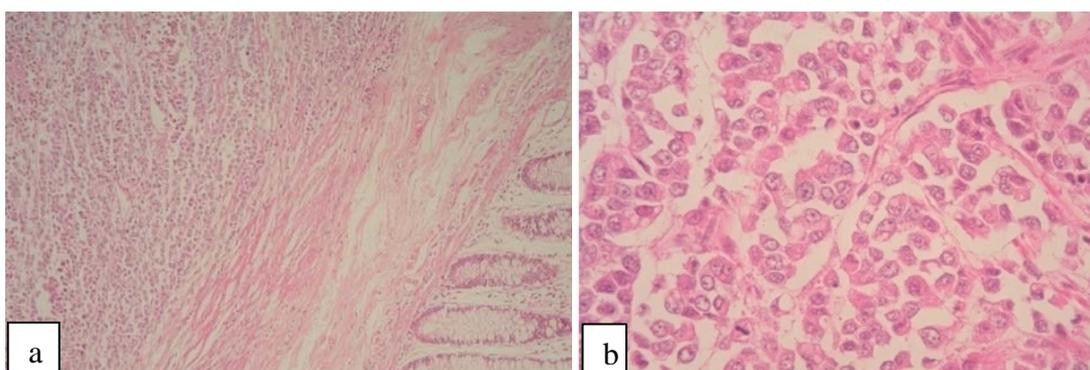


Figure 2 (a). Microscopic examination showed tumor cells infiltrating between the overlying mucosa (H&E, x100)
(b). Higher view of a (H&E x400).

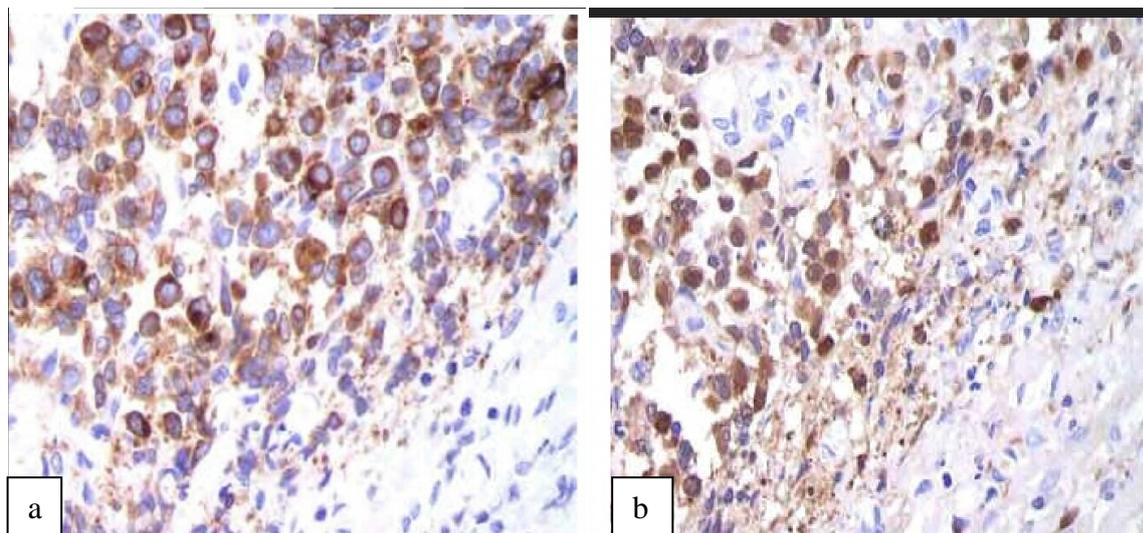


Figure 3. (a&b) Immunohistochemical staining of the tumor cells with melan A/MART-1 (a), and with S100 (b) (x400)

Discussion

Malignant melanoma of the rectum is rare and has very poor prognosis. The incidence has been reported to be 0.4-3% of all malignant melanoma and 0.1-4.6% of all anorectal malignant tumors^(4,5). Melanomas of the anorectum are the most common after melanomas of the skin and retina. Moore was the first person to report melanoma of the anus and rectum in 1857⁽¹²⁾. Malignant melanomas occur frequently in the anorectum because of the presence of abundant melanocytes in the mucosa of the anal canal⁽⁶⁾. Usually affects women in the fifth or sixth decade⁽¹³⁾, and presents with rectal bleeding or altered bowel habits⁽⁵⁻⁶⁾. The absence of early clinical manifestations and lack of clinical suspicion contribute for delayed diagnosis. Up to 60% of the cases have metastasis at the time of diagnosis⁽⁷⁻¹⁰⁾. In our case the disease was

limited to rectum and anal canal with two regional lymph nodes. The factors for poor prognosis includes, advanced disease at the time of the diagnosis and rich vascularity which increases the risk of hematogenous metastasis⁽⁸⁻¹⁵⁾. Abdomino-perineal resection is the treatment of choice for patients with <2mm wide lesion⁽⁸⁻⁹⁾. Radiotherapy is palliative in locally extensive tumors while combined with chemotherapy is used for metastasis⁽¹¹⁾.

Conclusion

Although anorectal melanomas are rare, this should be considered as one of the differential diagnosis in malignancies other than adenocarcinoma. So early diagnosis with histological features confirmed with immunohistochemistry will help in appropriate management and long term survival of the patient.

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تقرير حالة ورم الخلايا القتامينية الخبيث الابتدائي في الناحية المستقيمية - الشرجية

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الملخص

يعدُّ الورم القتاميني الخبيث (ميلانوما) في الناحية المستقيمية - الشرجية من الأورام النادرة جداً حيث تتراوح نسبة حدوثه ما بين 0.4-1% من مجمل الأورام القتامينية الخبيثة وهي تتصف بسوء الإنذار بحيث تتراوح نسبة البقيا ما بين 8-23 شهرا. ويتضمن علاج هذا الورم الخبيث كل من التداخل الجراحي والعلاج الشعاعي والعلاج الكيميائي. الحالة المقدمة هنا هي لمريضة تبلغ من العمر 75 سنة تشكو من نزف دموي خلال الشرح منذ خمسة أشهر. وعند إجراء التنظير الهضمي السفلي تبين وجود نمو نابت بقياس 5سم، وقد أخذ منه خزعات متعددة، وتبين مجهريا وجود خلايا خبيثة في المنطقة تحت المخاطية. وأجري استئصال للشرح عبر البطن. وقد أظهرت الدراسة النسيجية الإصابة بورم الخلايا القتامينية الخبيث في المنطقة المستقيمة الشرجية. إن الكشف المبكر والعلاج لهذا الورم عالي الخباثة سيزيد من فترة البقيا وتحسين الحالة العامة للمريض.

الكلمات الدالة: ورم الخلايا القتامينية الخبيث الابتدائي في الناحية المستقيمية-الشرجية، استئصال ما حول الشرح عبر البطن.