

# Anorectal amelanocytic malignant melanoma: Case report

## Anorektal amelanositik malign melanom: Olgu sunumu

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**Received/Accepted:** October 31, 2016 / May 22, 2017

**Conflict of interest:** There is not a conflict of interest.

### SUMMARY

Primary anal malignant melanoma is a rare tumor of the anal region. Usually it was misdiagnosed as hemorrhoids or anal polyp so delayed diagnosis can lead complicated status of the disease. Clinical and histopathological diagnosis may be difficult due to uncommon presentation and similar morphological findings when compared with other malignant anal tumors. Effectiveness of the chemotherapy and radiotherapy are limited. Surgical resection is still seen as standart treatment option and abdominoperineal resection (APR) and wide local excision are the most preferred methods. In this paper we presented 55 year-old male patient with recurrent anal malignant melanoma treated with abdominoperineal resection.

**Keywords:** Melanoma, anorectum, cancer

### ÖZET

Primer anal malign melanom, anal bölgenin nadir görülen bir patolojisidir. Genellikle anal melanomaya hemoroid ya da anal polip olarak yanlış tanı konulmakta, bununla birlikte tanı ve tedavideki bu gecikme hastalığın komplike bir tablo ile karşımıza çıkmasına neden olmaktadır. Kemoterapi ve radyoterapinin yararlılığı sınırlıdır. Cerrahi standart tedavi yöntemi olup abdominoperineal rezeksiyon (APR) veya genişletilmiş lokal eksizyon en sık tercih edilen yöntemlerdir. Biz bu yazıda 55 yaşında APR ameliyatı yapılan nüks anorektal malign melanomu olan erkek hastayı sunmayı amaçladık.

**Anahtar sözcükler:** Melanom, anorektum, kanser

### INTRODUCTION

Primary anal malignant melanoma is a rare tumor of the anal region<sup>1,2</sup>. Usually it was misdiagnosed as hemorrhoids or anal polyp so delayed diagnosis can lead complicated status of the disease<sup>3</sup>. Anal malignant melanoma has a very poor prognosis and is accounted for about 0,8-1% and 0,7 % of all anorectal malignancies and all malignant melanomas respectively. It usually tends to present in the fifth to sixth decade of life and male to female ratio is equal. Clinical and histopathological diagnosis may be difficult due to uncommon

presentation and similar morphological findings when compared with other malignant anal tumors<sup>4</sup>. Effectiveness of the chemotherapy and radiotherapy are limited. Surgical resection is still seen as standart treatment option and abdominoperineal resection (APR) and wide local excision are the most preferred methods<sup>5</sup>. Regardless of the wideness of the resection, local and distant organ spread is still high and the 5-year survival rates are ranged from 10%-20%<sup>6,7</sup>. In this paper we presented 55 year-old male patient with

recurrent anal malignant melanoma treated with abdominoperineal resection.

### CASE REPORT

55 year old male patient was admitted to the surgical department with pain and anal mass. After physical examination the anal polyp was removed with local excision. Final histopathological diagnosis was anorectal malignant melanoma and the surgical margins were positive. PET/CT imaging revealed pathological involvement in the left inguinal region (SUV max:3.4).(Figure 1-2). The patient had underwent surgery and extended local excision with left inguinal lymph node dissection was done. The surgical margins were negative and no positive lymph node was detected at the

pathological specimen. After 1-year follow-up an anorectal mass was detected in colonoscopy and multipl biopsies was taken. Histopathological examination of the biopsies were determined as the recurrent malignant melanoma. PET/CT imaging revealed a malignant rectal mass narrowed the lumen and extended through the anal canal measured as 5,5 cm. There were no distant organ metastasis. After preoperative preparation APR was performed due to extension of the mass through the anal canal and surrounding tissue. Rectal amelanocytic malignant melanoma was reported in final histopathological examination of the specimen (Figure 4-5). The patient was discharged on fifth day postoperatively without any complication.

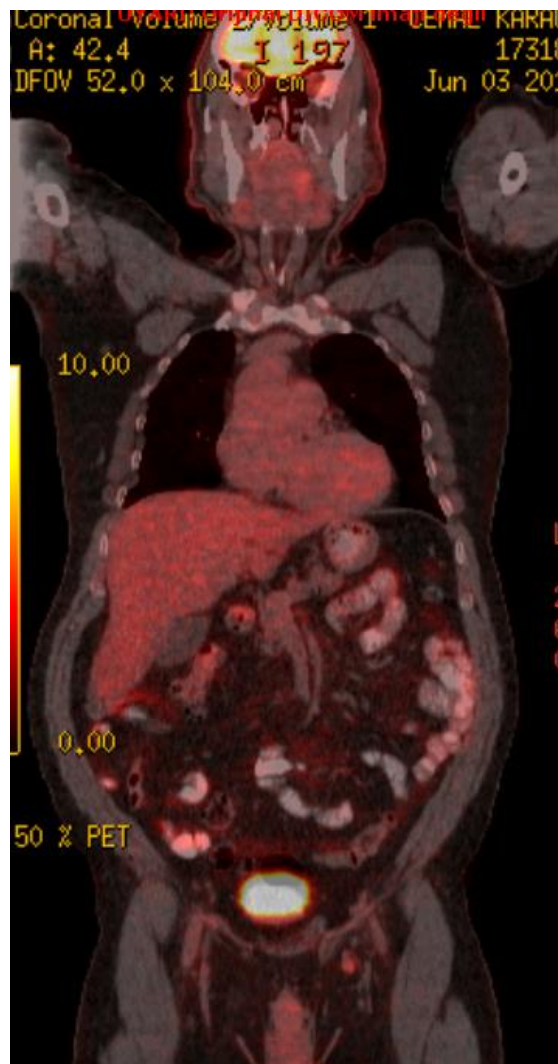


Figure 1

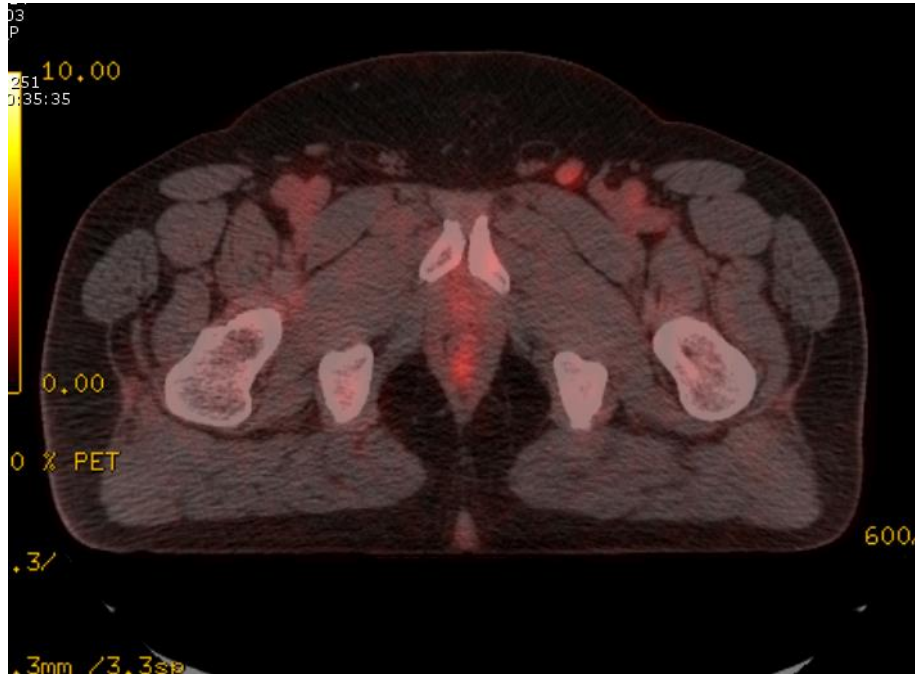


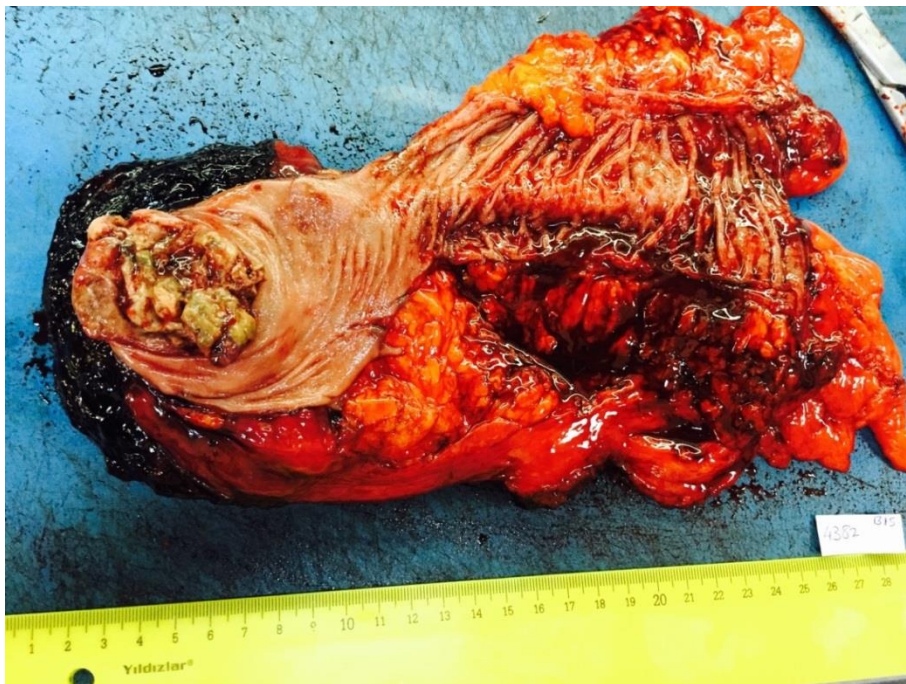
Figure 2



Figure 3



**Figure 4**



**Figure 5**

## **DISCUSSION**

Primary malignant melanoma of the gastrointestinal system is quite rare. malignant melanomas are very aggressive tumors. After skin and eye, the third most common location of this disease is anorectal region<sup>3</sup>. First primary malignant melanoma was published by Moore et al. at 1857 after that nearly 500 cases were reported in published literature<sup>8,9</sup>. Although adenocarcinomas and squamous cell carcinomas are the most common tumors of the colorectal and the anal canal

neoplasms, some neoplasms include lymphomas, melanomas, Kaposi's sarcoma and leiomyosarcomas can be seen<sup>8-10</sup>. Usually it arises from mucosa near the Dentate line, and shows itself as a polypoid lesion around the anorectal ring. Macroscopically it is difficult to distinguish from hemorrhoids or polyps. Anorectal malignant melanomas can arise from melanocytes in the non-keratinized stratified squamous epithelium or transitional epithelium below the dentate line. Malignant melanomas arise from the melanomas

settled in the rectum mucosa are very very rare. The most common symptom is rectal bleeding so it can be misdiagnosed as hemorrhoid. Other symptoms are palpable mass, pain, altered bowel habits, pruritus ani, tenesmus. Our patient had a similar symptoms reported in literature and was treated firstly as anal polyp.<sup>3</sup> It is very important to use modern imaging methods include abdomen and pelvic computerized-tomography, endoluminal ultrasonography, magnetic resonance imaging of the pelvic and whole body PET/CT with a good history and physical examination in cases diagnosed by histopathological examination, because the most important factor to predict the survival is the stage of the disease<sup>11</sup>. Anorectal malignant melanomas usually don't cause obstruction because they grow up longitudinally. Some cases presented with palpable inguinal lymph nodes, other regional lymph node or distant organ metastasis. About 40%-70% of the patients, there were regional or distant lymph node metastasis at the diagnosis. Because of the complexity and intensity of the regional lymph node flow, inguinal and mesenteric lymph node involvement can be seen. Today PET/CT scan often used for determine the distant organ metastasis or lymph node involvement. We used PET/CT for this purpose and performed left inguinal lymph node dissection due to pathological involvement in the left inguinal region, however, there were no metastatic lymph node among the fourteen lymph node dissected. (figure 1-2). Although surgery is the certain treatment of the disease, there are no agreement for the surgical methods in the literature. Some authors suggest APR especially for localized small tumors, however, opposites suggest sphincter preserving wide local excision because many of the patients died due to diffuse systemic disease or distant organ metastasis<sup>12</sup>. Early studies in the past suggested with aggressive APR and more wide negative surgical board, better survival rates and lower local recurrence rates would be obtained and doing lymphadenectomy controlled lymphatic spread<sup>9,12</sup>. In a 85 cases serie, authors recommended APR because of higher survival rate between 1929-1993, at Memorial Sloan-Kettering Cancer Research Center<sup>13</sup>. However the same center reported that there were no difference with disease free and overall survival rates between APR and 4x5x2"1 local excision in a serie of 64 cases between 1984-2003. The advantages of the local excision are short operation time, fast wound healing, not requiring stoma, short hospitalization, fast recover of the bowel functions. However some authors reported higher local recurrence rates with wide local excision<sup>14</sup>.

Although radical surgery is necessary in large tumors wide local excision is recommended in technically possible cases. In our patient firstly wide local excision was performed, in follow up recurrence was detected in anal canal. In PET/CT there were no surrounding tissue involvement but there were some lymph nodes, so APR was done.

Malignant melanomas are seen thirdly anorectal region after skin and eye. Primary anorectal malignant melanomas arise from melanocytes in the anal canal<sup>15</sup>. The etiology of the amelanocytic is still unknown. Pigment loss in the melanocytes due to abnormal melanogenesis or agenesis of the melanosomes can occur. Also in the amelanocytic melanoma cases the loss of tyrosinase enzyme which is necessary for the synthesis of melanin are suggested, otherwise some authors suggested that because of melanin production is very low concentration, it can't be detected clinically and histopathologically<sup>16,17</sup>. In our patient the melanoma is amelanocytic.

**In conclusion**, anorectal malignant melanoma is a rare condition. If a patient suffers recurring rectal bleeding, pain anorectal malignant melanoma must be considered as a differential diagnosis. Unfortunately, delayed diagnosis cause increased morbidity and mortality. Despite the poor prognosis, APR and wide local excision are used as a treatment choice.

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