



A Rare Case Report: Anal Malignant Melanoma and Evaluation of the Literature

Nadir Karşılaşılan Bir Olgu: Anal Malign Melanom, ve Literatürün Gözden Geçirilmesi

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Abstract

Anal malignant melanomas are relatively rare tumors. Because of their polypoid structure, they can be confused with benign anorectal diseases. Wide excision and lymph node dissection is recommended especially in melanomas including lymph node metastasis because it reduces local recurrence. We performed abdominoperineal resection + bilateral inguinal + bilateral iliac + paraaortic lymph node dissection in our 77-year-old female patient with the diagnosis of anal malignant melanoma as a result of colonoscopic biopsy performed on the polypoid mass in the anal canal. Since 2 lymph nodes located outside the pararectal fascia persisted in postoperative MRI, we performed lymph node excision with a posterior approach. And the pathology result of the excision performed in this second session was found to be compatible with metastasis. Lymph node dissections are beneficial in terms of local recurrence and survival, especially in patients with anal malignant melanoma with lymph node metastases.

Keywords: Anal Malignant Melanoma, Anal Cancer, Anal Malignancy

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Öz

Anal malign melanolar görece nadir tümörlerdir. Sıklıkla polipoid yapıda olmaları nedeniyle benign anorektal hastalıklarla karıştırılmaktadır. Özellikle lenf nodu metastazını içeren melanomlarda geniş eksizyon ve lenf nodu diseksiyonu lokal nüksü azaltması nedeniyle önerilmektedir. 77 yaşındaki kadın hastamıza anal girimdeki polipoid kitleye yapılan kolonoskopik biyopsi sonucunda gelen anal malign melanoma tanısı ile abdominoperineal rezeksiyon + bilateral inguinal + bilateral iliyak + paraaortik lenf nodu diseksiyonu yapıldı. Pararektal fasyanın dışında yer alan iki adet lenf nodunun da postoperatif MR da sebat etmesi üzerine posterior yaklaşımla lenf nodu eksizyonu yapıldı ve bu ikinci seanssta yapılan eksizyonun patoloji sonucu metastazla uyumlu görüldü. Özellikle lenf nodu metastazı olan anal malign melanom hastalarında lenf nodu diseksiyonları lokal nüks ve sağkalım açısından faydalıdır.

Anahtar Kelimeler: Anal Malign Melanom, Anal Kanser, Anal Malignansiler

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Introduction

Anal malignant melanoma is an extremely aggressive and luckily very rare tumor. Surgical resection is the main treatment, unfortunately value of adjuvant treatments is limited (1). Only 0.5–2% of anorectal cancers and less than 2% of melanomas are caused by it. The most common site of primary gastrointestinal melanoma is the anal canal. Only 2% of cutaneous melanomas metastasize to gastrointestinal tract, and only 2% of these metastases are rectal metastases (1). The median age at which anorectal melanoma first appears is 55, but this range is broad (2).

All melanomas develop from melanocytes. When melanocytes are exposed to carcinogenic stimuli, malignant transformation into melanoma is observed. It has been clearly demonstrated that risk of cutaneous melanoma increases with exposure to sunlight, especially ultraviolet B radiation. It is unclear what triggers the development, though, as the anal mucosa is not exposed to light (3).

Melanin pigment in malignant cells is the basis of histological diagnosis. Presence of junctional changes helps to establish the diagnosis. Without melanin or junctional changes, immunohistochemistry may be helpful. S-100, HMB-45 and vimentin stained positively respectively in 78, 94 and 100% of the tumors(4). The aim of our case report is to present a patient with primary anal malignant melanoma who underwent abdominoperineal resection + radical lymph node dissection in our clinic.

Case Report

A 77-year-old female patient was admitted to the outpatient clinic of an external center with complaints of swelling, bleeding and pain in the rectal region. First evaluation of the patient was in favor of hemorrhoidal disease and medical treatment was started. Afterwards, the patient was admitted to our general surgery outpatient clinic because her complaints did not regress despite medical treatment. On rectal examination, a stalked polyp of approximately 5 cm in diameter, which almost completely covered the lumen, was palpated at the anal entrance. In the colonoscopy, multiple biopsies were performed from the polypoid lesion at the anal entrance (Figure 1, 2)



Figure 1. Polypoid mass closing the lumen in colonoscopy.



Figure 2. Polypoid mass closing the lumen in colonoscopy.

Pathological evaluation was reported as "In sections, it does not contain surface epithelium, forms layers and islands on the background of necrotic tissue, has large pleomorphic nuclei, prominent eosinophilic nucleoli, shows high mitotic activity, positive with HMB45, S100, MelanA and Vimentin, p16, CK7, CK20, CDX2, ER, PR, CEA. Negatively stained neoplastic proliferation with LCA, PanCK, EMA, SMA was observed. Malignant melanoma was considered.". Ki 67 proliferation index was found around 35%.

Thereupon, the patient was examined with imaging methods in terms of metastatic disease and the MR imaging of the patient was reported as " A polypoid mass lesion filling the lumen is observed in the 5 cm distal part of the rectum. Heterogeneous diffusion features and enhancements are observed in the lesion. As far as can be seen, the lesion extends proximal to the anal canal. The 11x8 mm nodular lesion on the right lateral wall of the pelvis with diffusion restriction and enhancement is compatible with metastatic LAP. In addition, another 10x8 mm metastatic lymph node is observed on the right lateral wall of the pelvis, close to the acetabular roof. No significant metastatic lesion was observed in the perirectal adipose tissue. The mesorectal fascia is free." (Figure 3)

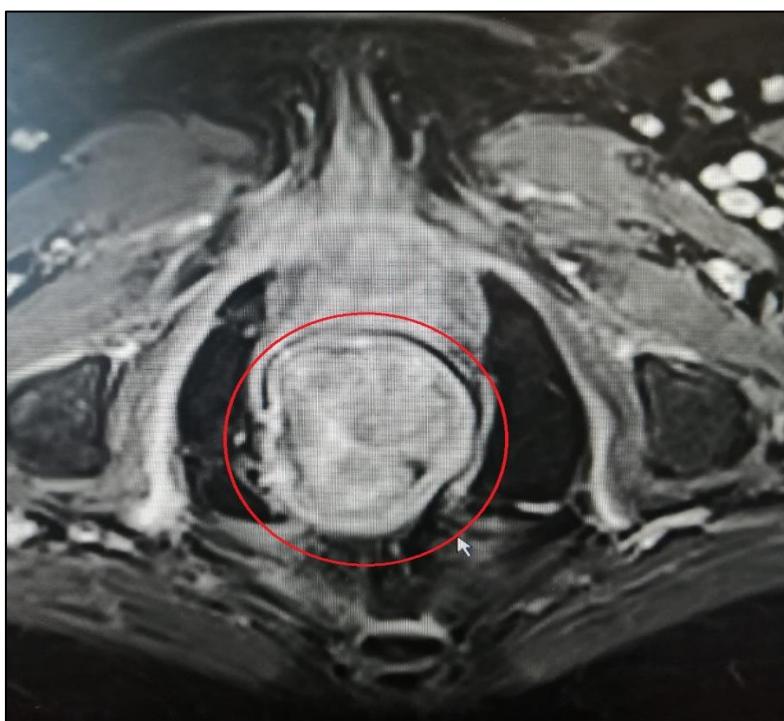


Figure 3. Malignant melanoma with polypoid appearance at the anal entrance on preoperative MRI.

The tumor board assessed the patient and made the decision to operate as abdominoperineal resection (APR), bilateral iliac, inguinal and paraaortic lymph node dissection. Afterwards, the patient underwent surgery.

The final pathology result of the patient was reported as; "The tumor is situated on the dentate line of the anal canal, 6x5x4.5 cm in size, shows polypoid development, invades the muscular layer of the intestinal wall, and is 2.5 cm from the nearest (distal surgical margin) surgical margin. There is perineural invasion, no lymphovascular invasion. Surgical margins are intact. Additionally, there is no tumor in the radial surgical margin. There were 5 paraaortic, 13 right pelvic, 7 left pelvic, 4 left inguinal, 6 inguinal lymph nodes from both right and left sides which was sent for frozen diagnosis, and 15 lymph nodes from the intestinal periphery. Overall 50 lymph nodes were reactive."

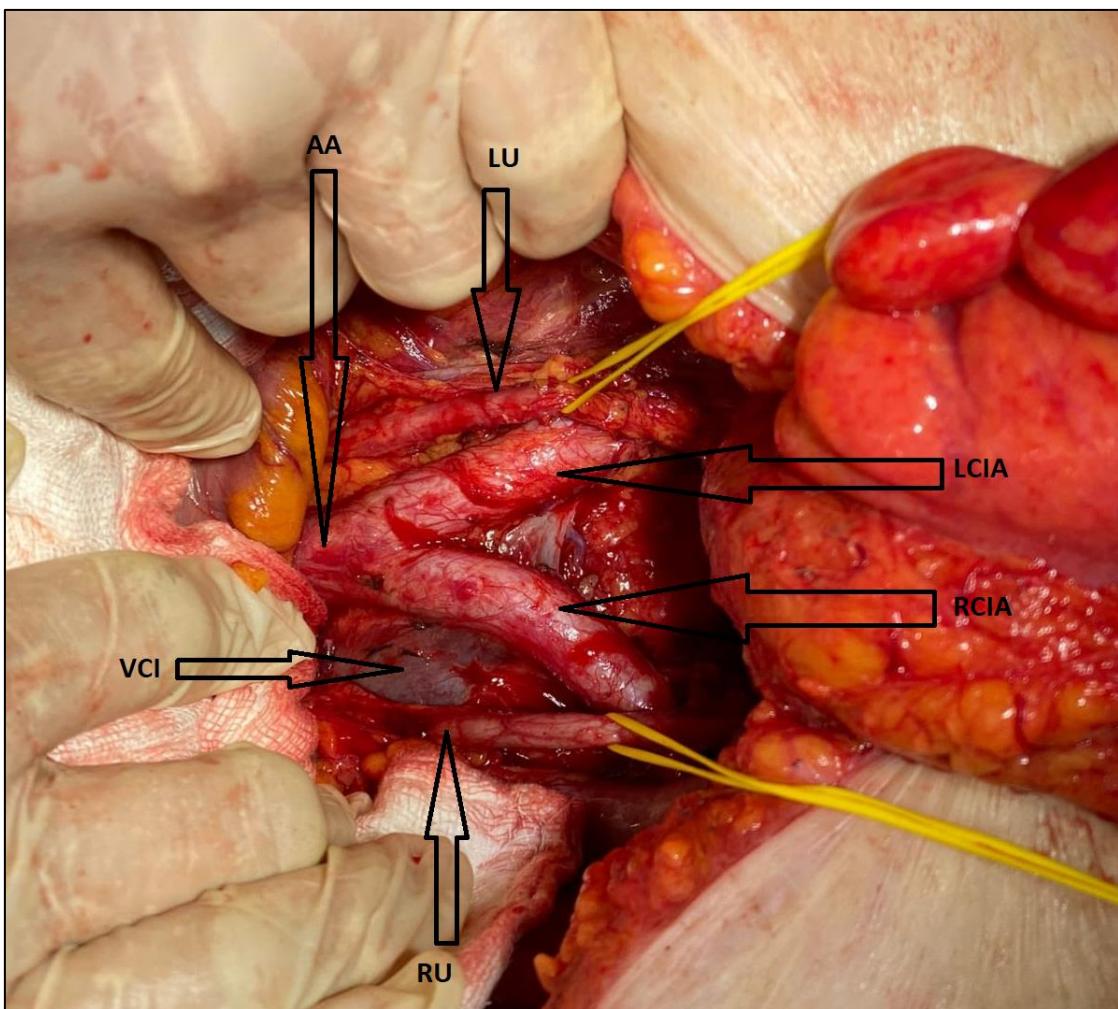


Figure 4. Operation image of abdominoperineal resection + bilateral parailiac + paraaortic lymph node dissection (AA: abdominal aorta, LU: left ureter, RU: right ureter, LCIA: left common iliac artery, RCIA: right common iliac artery, VCI: vena cava inferior).

For the patient's postoperative assessment, a new contrast-enhanced MRI was taken. It was observed during the radiological evaluation that the lymph node with pathological appearance, which was described outside the pararectal area in the previous MRI, on the right pelvic wall close to the acetabular roof, was still persistent. Thereupon, transsacral lymph node dissection with stereotaxic marking was planned for the patient (Figure 5).



Figure 5. CT image of 2 pathological lymph nodes and stereotaxy wire at the level of the sacrococcygeal ligament.

With the use of intraoperative ultrasound, the lymph nodes were removed during the surgery. On the postoperative 2nd day, intestinal contents were collected through patient's drain placed in the pelvic. The patient underwent an emergency laparotomy due to concerns of an intraoperative injury. It was observed that the ileal loop located in the pelvic fossa during the exploration was perforated with a width of 3 mm in the second operation due to secondary thermal damage to the energy devices used during the exploration. This region underwent wedge resection and anastomosis. The lymph nodes excised during the second procedure were shown to be consistent with metastasis in the final pathological assessment.

Discussion

Approximately 8300 new cases of anal cancers are diagnosed causing approximately 1200 deaths annually. It is a very uncommon malignancy that makes up around 3% of all gastrointestinal malignancies (5, 6).

Surgical excision remains primary treatment. Surgery is the preferred form of therapy; however, the best method is not universally agreed upon. (7,8). Since metastatic lymph nodes were observed in the MR imaging of our patient, APR and paraaortic + parailiac + bilateral inguinal lymph node dissection was preferred.

Chemotherapy and radiation have a restricted role. Although the disease is well controlled with WLE and adjuvant radiotherapy, it has been stated by some authors that the general prognosis of the disease is not affected (9).

Despite the addition of new treatment modalities over time, the lack of improvement in survival for the last 30 years indicates the requirement for treatment that prioritizes quality of life, in a multidisciplinary setting (8).

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