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MELANOMA OF THE ANAL CANAL WITH MELANOSIS OF THE PERITONEUM - CASE REPORT

Dragoslav Miljković, Vanja Pecić, Vanja Pecić, Milica Nestorović, Dragan Mihajlović, Branko Branković, Milan Radojković, Ljiljana Jeremić, Sunčica Mihajlović and Dejan Petrović

Primary malignant melanoma of the anorectum is rare. It accounts for 0.2-0.3% of all malignant melanomas. Prognosis is very poor since majority of patients dies within first two years.

We report a case of a 76 year-old man with anoretal melanoma. Digital rectal examination revealed a mass on the anterior wall. Biopsy report favored two possibilities, first, malignant melanoma and second, poorly differentiated carcinoma. Abdominal ultrasonography and chest X-ray did not show signs of distant metastasis. Computed tomography of abdomen and pelvis revealed presence of a large, irregular hypodense mass with heterogeneous enhancement in anal canal accompanied by enlarged lymph nodes. Abdomino-perineal resection was performed. Histopathological examination of resected specimen confirmed diagnosis of malignant melanoma. Postoperative course was uneventful. However, patient died one year after due to metastasis.

Surgery is the mainstay of curative treatment of anorectal melanoma, effective systemic adjuvant therapy has been lacking. Long-term survival is rare, as most patients die of disseminated systemic disease. *Acta Medica Medianae 2011; 50(4):45-48.*

Key words: *malignant melanoma, anorectum, abdominoperineal resection*

Clinic for General Surgery, Clinical Center Niš, Niš, Serbia

Contact: Milica Nestorović
Clinic for General Surgery, Clinical Centre Niš
Bul. Zorana Đinđića 48, 18000 Niš, Serbia
E-mail: milica20@yahoo.com

Introduction

Primary malignant melanoma of the rectum is an exceptionally rare neoplasm with documentation limited to case reports over many years and literature reviews thereafter (1,2). Over the years, many cases have been disputed on the grounds of proof of local origin with extension from an anal lesion often believe to be the primary site (1). Moore was the first person to report melanoma of the anus and rectum in 1857 (3). Anorectal malignant melanoma accounts for 1-1.5% (4) of all malignant melanoma cases, and 0.1%-4.6% of malignant tumors of the rectum and anus (5). Prognosis is very poor with a median survival of 24 months and a 5-year survival of 10% (2,6). Almost all patients die of metastases (7). We observed a patient with progressive generalized hyper pigmentation of the peritoneum caused by an anorectal melanoma.

Case report

A 76 year-old man presented with history of bleeding per rectum in 4 months duration. Blood was mostly seen mixed up with stools with occasional passage of mucus. He complained about generalized weakness and sudden weight loss in the past 2 months. In past medical history he reports no previous hospitalizations. He suffers from hypertension, takes medications on regularly basis. He also had grade II hemorrhoids, which had bled two years back. He did not smoke or drink. Family history was negative for large intestine malignancy.

General examination revealed marked pallor of skin with many nevi on the body, size 4-8mm (Figure 1). There were no palpable lymph nodes. There was no palpable abdominal organomegaly. Digital- rectal examination revealed a mass, 5 cm in diameter, on the anterior wall of the rectum, 3 cm from the anal verge. Rectosigmoidoscopy demonstrated a bluish-maroon proliferate mass lesion in the rectum involving half the circumference. Biopsy was performed and histopathological report favored two possibilities, malignant melanoma and poorly differentiated carcinoma.

The results of complete blood chemistries and blood count were within normal range, except lower red blood cell count. Abdominal ultrasonography showed no evidence of metastases. X-ray of the chest did not show evidence of pulmonary metastasis or any other finding. Computed tomography of abdomen and pelvis revealed presence of a large, irregular hypodense mass with heterogeneous enhancement in the rectum. Few enlarged pararectal lymph nodes were present.

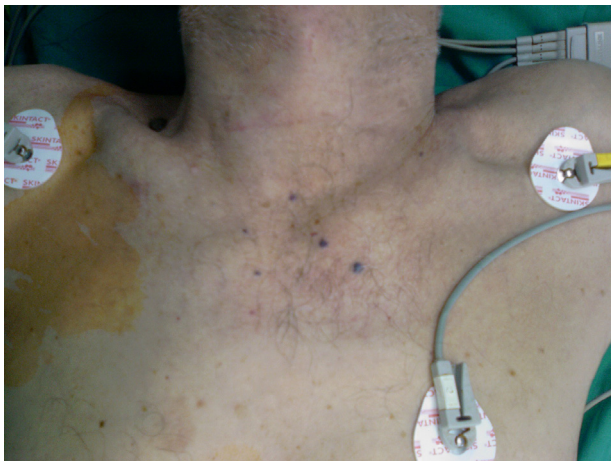

Figure 1. Multiple nevi on the body

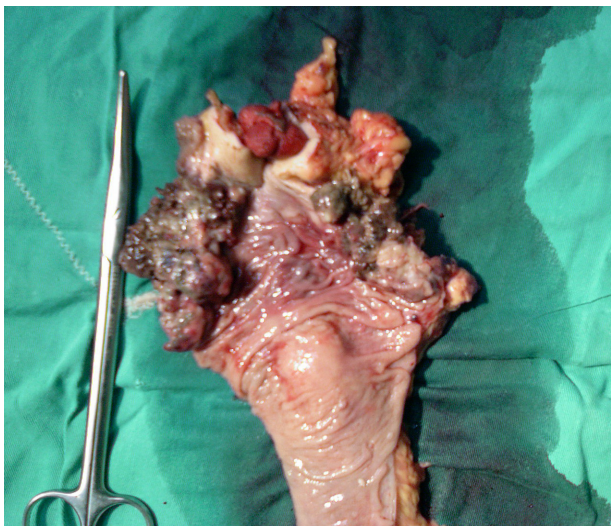

Figure 2. Specimen after abdomino-perineal resection

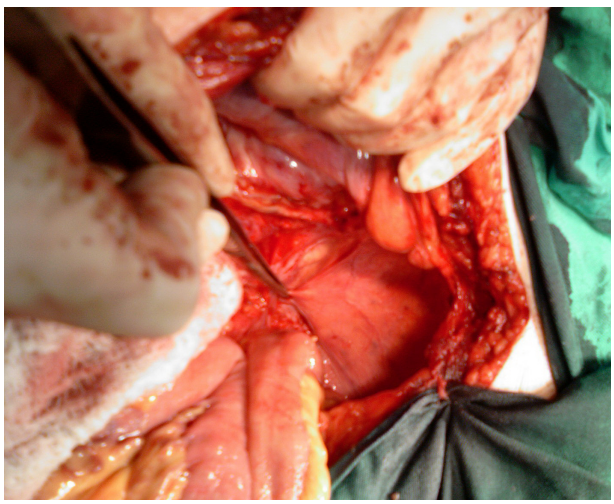

Figure 3. Melanosis of the peritoneum

Patient was preoperatively prepared and abdomino-perineal resection was performed (Figure 2). During the operation, following abdominal exploration, multiple brown and black dots were recorded on the peritoneal peritoneum (Figure 3). Postoperative course was uneventful and was discharged on the seventh postoperative day. Histopathology of the resected specimen revealed malignant melanoma of the rectum (S-100 stain positive) with complete rectal wall infiltration and metastatic lymph nodes and melanosis of the peritoneum. Patient was referred to adjuvant chemotherapy and the disease seemed to be in remission. One year after the initial diagnosis, however, patient died of intra-abdominal metastases.

Discussion

Etiology of melanoma has been linked to exposure to UV-B rays. Cutaneous melanoma is strongly linked to UV-B rays and is particularly related to sun blistering events in the childhood (8). Occurrence of melanoma in locations like rectum where the "sun never shines" is still an ambiguity (9). Genaralised melanosis or peritoneal melanosis caused by rectal melanoma are extreamly rare. The pathogenesis of this phenomenon is not well understood. The typical slate-gray discoloration of generalized melanosis was explained in 1954 by Fitzpatrick et al. to be the result of oxidation of melanin precursors that escaped from the tumor into the blood. These intermediates cross the capillary membrane and are converted into melanin by oxidizing systems within the histiocytes and possibly in the extracellular fluids of the dermis (10). According to some authors this phenomena is analogous to the „melanosis" of squamous mucosal membranes (nasal or oral cavity, anus, vagina, esophagus) commonly seen in the proximity of primary melanomas at those sites (11).

Patients with malignant melanoma of rectal localization generally present with rectal bleeding and a sensation of a mass which is usually attributed to hemorrhoids, as in this case, or to presence of polyp. Ignoring or misdiagnosis leads to diagnosis in advanced stage of disease (12). At this moment there is no consensus on which threatment approach is favourable. In most published papers, surgery is the mainstay of curative treatment of anorectal melanoma. The main determinant of prognosis like in skin melanoma is the tumor thickness (13). The surgical procedure of choice ranges from an abdomino-perineal resection to local excision with or without adjuvant radiotherapy (2).

The effect of adjuvant chemotherapy and postoperative radiotherapy on the prognosis is a controversial topic and further studies are needed to demonstrate the benefits of these treatment modalities (12). The field of melanoma therapeutics has experienced a dramatic change through recent discoveries of novel therapeutics targeting known oncogenes and immunotherapeutic antibodies. Immuno-therapeutic options have diversified, creating opportunities not limited strictly to the inpatient setting (14).

Long-term survival is rare, as most patients die of disseminated systemic disease regardless of treatment; for this reason, some authors do not consider extensive radical surgery to be the treatment of choice and prefer wide local excision

as initial treatment (15). Only in case of large obstructive tumors one should perform abdomino perineal resection (2).

The factors that may account for the poor prognosis include the advanced nature of the disease when diagnosed, ulceration, the rich vascularity of the rectal mucosa, heightened risk of hematogenous metastasis, and the probable high biological aggressiveness of the tumors (16).

In conclusion, a malignant melanoma of the anorectum is a rare disorder. The prognosis is poor and patients often have already disseminations at the time of diagnosis. A standard approach to managing has not been established since most data is based on case reports and retrospective analysis. Hopefully, new modalities of biological or immunological therapies will improve survival of these patients.

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MELANOM ANOREKTUMA SA MELANOZOM PERITONEUMA - PRIKAZ BOLESNIKA

Dragoslav Miljković, Vanja Pecić, Milica Nestorović, Dragan Mihajlović, Branko Branković, Milan Radojković, Ljiljana Jeremić, Sunčica Mihajlović i Dejan Petrović

Primarni maligni melanom anorektuma je redak. Čini svega 0.2-0.3% svih malignih melanoma. Prognoza je loša. Većina bolesnika umre u prve dve godine od postavljanja dijagnoze.

Prikazan je sedamdesetšestogodišnji muškac sa anorektalnim malignim melanomom. Rektalnim tušeom nađena je palpabilna masa na prednjem zidu rektuma. Nalaz biopsije ukazivao je na dve mogućnosti, prvu, maligni melanom i drugu, slabo diferentovani karcinom. Ultrazvukom abdomena i radiografijom grudnog koša nije utvrđeno postojanje udaljenih metastaza. Kompjuterizovana tomografija abdomena i karlice potvrdila je prisustvo hipodenzne mase uz uvećane limfne noduse pararektalno.

Bolesnik je operisan, urađena je abdomino-perinealna resekcija po Miles-u. Histopatološkim pregledom preparata potvrđeno je da se radi o malignom melanomu. Postoperativni tok je protekao uredno. Ipak, bolesnik umire godinu dana nakon postavljanja dijagnoze usled diseminacije primarnog procesa.

Hirurški tretman je stub lečenja malignog melanoma anorektalne regije. Nedostaje efikasna sistemska adjuvantna terapija. Dugo preživljavanje je retkost, većina bolesnika umire usled metastatskog procesa. *Acta Medica Medianae 2011;50(4):45-48.*

Ključne reči: *maligni melanom, anorektum, abdominoperinealna resekcija*