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Malignant rectal melanoma

Case report

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Malignant rectal melanoma. Case report

AIM: *The aim of this study was to report a case concerning diagnostic and management of Anorectal melanoma.*
MATERIAL: *A 71 years old white man presented in our Institute with anal pain, tenesmus, blooding rectal during the last five months. The ano-rectal examination showed a brownish painful mass in the anal canal. The colonscopy and endoscopy showed a big stenotic mass from anal canal to medium rectum with a diameter of approximately 90 mm.*
RESULTS: *Biopsy of the rectal mass was performed and the histopatological examination showed malignant ephitelioid cells, pigmented melanoma. The patient was treated by abdominoperineal resection with dissection of lymphnodes. Result of histopatological examination was ulcerated Malignant melanoma of the anal canal, growing polypoid with spindle cells and epithelioid infiltrating the mucosa, submucosa and the internal sphincter muscle. Vascular invasion. Subtotal lymph node metastasis in 3 of 17.*
DISCUSSION: *Anorectal melanoma is an uncommon and aggressive disease. The anorectum is the third most common location of malignant melanoma after the skin and retina. Lesions are difficult to diagnose because many are amelanotic and patients present with nonspecific complaints such as anal discomfort or rectal bleeding.*
After diagnosis, the main treatment available is surgical resection. Sentinel lymph node mapping has an unclear role in its management. Adjuvant therapy has long been recommended; however, there are no strong data to support its use.
CONCLUSIONS: *There is no convincing evidence to indicate that abdominoperineal resection did not improve the survival rate of patients with malignant rectal melanoma as compared to the wide local excision, while the wide local excision had advantages in lower surgical risk and allowing patients to avoid permanent colostomy.*

KEY WORDS: Cancer, Rectal melanoma, Surgery

Introduction

Anorectal melanoma is an uncommon and aggressive disease. The anorectum is the third most common location of malignant melanoma after the skin and retina. The most common symptom is rectal bleeding, which is often mistaken for bleeding associated with hemorrhoids. Diagnosis is very difficult, and initial diagnosis may be incorrect in 80% of all cases. For patients with anorectal malignant melanoma, treatment strategy includes surgery, chemotherapy, and radiotherapy. However, the tumor tends to be considerably resistant to radiotherapy and shows a poor response to chemotherapy. The choice of wide local excision or abdominoperineal resection is also controversial. The prognosis is very poor, with less than 20% survival five years after diagnosis [1].

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Case Report

A 71 years old white man presented in our Institute with anal pain, tenesmus, blooding rectal during the last five months. He referred personal history of arterial hypertension, diabetes mellitus, atrial fibrillation and mitralic valvular disease. The ano-rectal examination showed a brownish painful mass in the anal canal (Fig. 1). The colonscopy and endoscopy showed a big stenotic mass from anal canal to medium rectum with a diameter of approximately 90 mm (Fig. 2-3). Biopsy of the rectal mass was performed and histopatologically examination showed malignant ephitelioid cells, pigmented melanoma. Computer tomography (CT) showed a thickening of the rectal wall and lymphonodes in the tissue adipose, in bilateral obturator district and bilateral lung metastasis (Fig. 4). Laboratory data as well as serum Ca 19-9 level was almost normal. The patient was treated by abdominoperineal resection with dissection of lympho nodes.

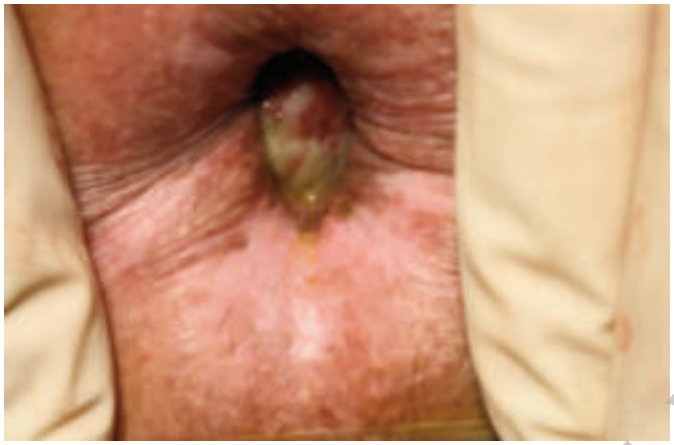
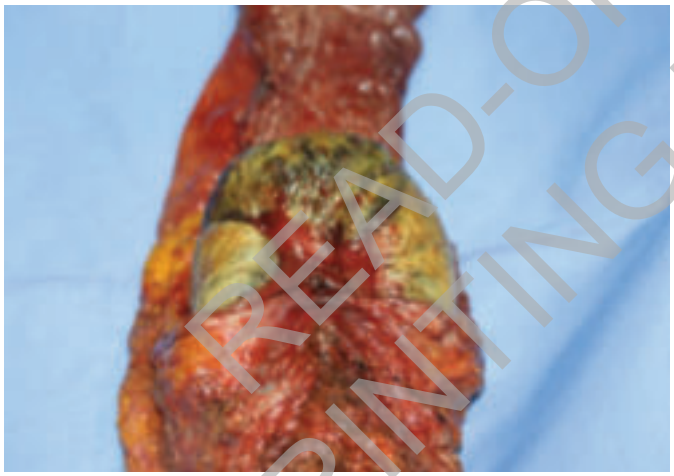


Fig. 1: Brownish painful mass in the anal canal.

Results

Result of histopatological examination was ulcerated Malignant melanoma of the anal canal, growing polypoid with spindle cells and epithelioid infiltrating the mucosa, submucosa and the internal sphincter muscle. Vascular invasion Subtotal lymph node metastasis in 3 of 17



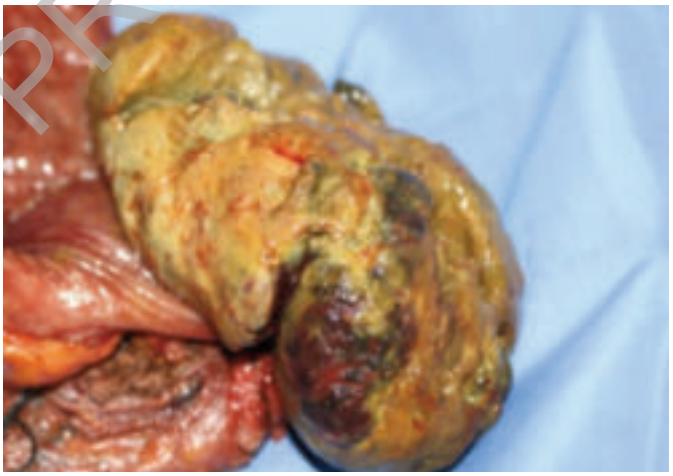


Fig. 2-3: Big stenotic mass from anal canal to medium rectum with a diameter of approximately 90 mm.

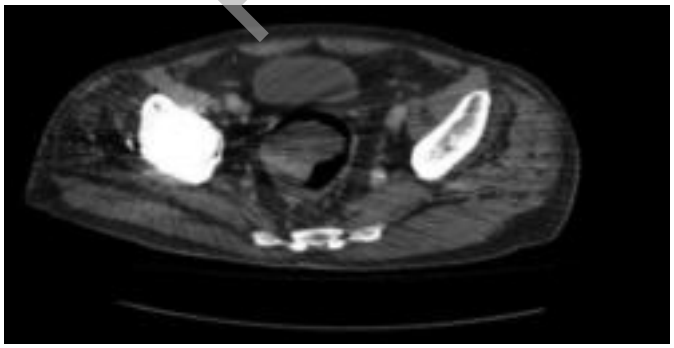


Fig. 4: Thickening of the rectal wall and lymphonodes in the tissue adipose in bilateral obturactor district.

Discussion and comments

Anatomic site is a predictive factor in subtypes of cutaneous and mucosal melanoma.

Determination of Anatomic Site of Origin

If Anatomic Location Documented
– Anal was defined as distal to the dentate line
– Anorectal was defined as at or traversing the dentate line
– Rectal was defined as proximal to the dentate line

If Exact Anatomic Location Uncertain
• Histology:
– Anal melanoma was identified by squamous mucosa
– Anorectal or Rectal melanoma was identified by colonic mucosa
• Lymphatic Drainage:
– Anal melanoma metastasized to inguinal lymph nodes
– Anorectal or Rectal melanoma metastasized to mesenteric lymph nodes

In a study at the Memorial Sloan-Kattering Cancer the authors examined the clinical relevance of location of origin of anorectal melanoma as a prognostic factor. Using a prospectively maintained database, clinical characteristics, management, and outcomes were compared according to site of origin. A retrospective review was conducted of patients diagnosed with anorectal melanoma from 1994-2010. Tumors were defined as anal, anorectal or rectal melanoma according to their anatomic relationship to the dentate line. Clinicopathologic factors were compared by Chi-square test. Time-to-event analysis was performed by Kaplan Meier analysis. Of the 96 patients included (41 anal, 32 anorectal, 23 rectal), patients with rectal and anorectal mucosal melanoma had advanced primary tumors (median Breslow thickness 12mm and 8mm respectively, p =0.002), while anal lesions could be found at earlier depths (median thickness 6.5mm). Patients with anal tumors more commonly underwent transanal excision (p <0.02) and sentinel lymph node biopsy (p=0.004) versus anorectal and rectal tumors. Patterns of recurrence were also distinct; nearly two-thirds of anorectal and rectal tumors recurred systemically, while anal melanoma more often recurred within the lymph nodes first (63%; p <0.02). Recurrence occurred in 24 (59%) patients with anal tumors, 23 (72%) anorectal tumors, and 16 (70%) rectal tumors. Median OS was 22 months for anal melanoma, 28 months for anorectal melanoma and 27 months for rectal melanoma. Recurrence and survival were not statistically different between the groups. This study represents the only series describing the outcomes of anorectal melanoma by anatomic location of the primary lesion. Lesions at or proximal to the dentate line present with more advanced disease, possibly related to a delay in diagnosis. Lesions distal to the dentate line more commonly recur within lymph nodes, which may represent differences in nodal drainage. Irrespective of location, long-term prognosis remains poor for all cases of anorectal mucosal melanoma; however, progress in the development of systemic therapy for this disease holds the promise for improved outcomes in the future [8].

Preoperative chemoradiotheraphy has a proven role in the treatment of various malignant diseases, particularly in locally advanced rectal cancer. The predominant role of preoperative CCRT is to reduce locoregional and pelvic cavity recurrence, and to obtain a higher rate of sphincter preservation via tumor shrinkage. In addition, it facilitates the removal of potential micrometastasis and lessens distant metastases [10].

Melanomas were previously known to be radio resistant. This was attributed to the ability of melanoma cells to repair sublethal and potentially lethal radiation damages. However, there is now adequate evidence to show that melanoma cells are radiosensitive. The use of adjuvant RT in mucosal melanoma has also increased.

Postoperative radiation therapy has been shown to improve loceregional control in head and neck mucosal melanoma and female genital melanoma. In anorectal melanoma, however, adjuvant radiotherapy has been used very sparingly. The use of hypofractionated radiotherapy has been associated with better response rates as compared to conventional fractionation in some series. The most important prognostic indicators in anorectal melanoma include stage of disease, size, duration of symptoms, nodal involvement and molecular markers like PCNA and Ki-67 and it is the invasion into the submucosa or deeper layers rather than the Breslow's thickness that determines the propensity for nodal or distant spread and ultimately, the survival in anorectal melanoma [11].

There has been a debate in the literature regarding the extent of surgery necessary for treatment of primary disease. Early studies have suggested that aggressive treatment of the primary anorectal lesion with abdominoperineal resection was associated with improved outcome, possibly due to regional lymphadenectomy. However, other studies, describing local excision of the primary anorectal lesion, without regional lymphadenectomy, have reported similar patterns of recurrence and survival with no significant increase in local failure. All studies are concordant with the fact that relapse is usually distant and lethal. There is no convincing evidence to indicate that radical resection of primary anorectal melanoma is associated with improvement in local control or survival. Patients with localized disease should undergo local excision whenever technically feasible. This approach aims to minimize morbidity and maximize quality of life in a disease that, even when localized, is rarely curable. The presence of tumor perineural invasion is an important prognostic factor and should be considered in future clinical trials [12].

The depth and size of tumor is one of the important prognostic factors. If the lesion is thick (>3 mm) and

large (>30 mm), curative surgery cannot be achieved [13,14]. In this case, conservative local excision and adjuvant therapy can result in a better prognosis. At the time of diagnosis, if anorectal malignant melanoma is already in the advanced stage, surgical options should be selected based on quality of life. For advanced stage, wide local excision with adjuvant radiotherapy and biochemotherapy could be done concerning favorable functional outcome and longer median survival. Recently, the use of immune-modulating agent for cancer treatment has increased, since chemotherapy or radiotherapy alone is insufficient to completely eradicate Malignant Rectal Melanoma. Moreover, immunotherapy can boost anticancer immunity. Immunochemotherapy is a chemotherapy regimen which includes an immunologic agent. Immunochemotherapy includes specific active immunization and adoptive immunotherapy based on antigenic system [13].

Conclusions

In conclusion, anorectal melanoma is a rare disease with a poor prognosis. Local, sphincter-sparing excision of the primary tumor followed by hypofractionated RT offers effective local therapy that is well tolerated. There is no convincing evidence to indicate that adbominoperineal resection did not improve the survival rate of patients with malignant rectal melanoma as compared to the wide local excision, while the wide local excision had advantages in lower surgical risk and allowing patients to avoid permanent colostomy. Patients with localized disease should undergo local excision whenever technically feasible. This approach aims to minimize morbidity and maximize quality of life in a disease that, even when localized, is rarely curable. Only when local excision of tumor mass was not possible technically, the abdominalperineal resection should be considered.

Riassunto

Il Melanoma Anorettale è una malattia rara e aggressiva ed è il terzo tipo più comune di melanoma maligno dopo quello della cute e della retina. Il sintomo più comune è il sanguinamento rettale, che è spesso scambiato per sanguinamento associato a emorroidi. La diagnosi è molto difficile, e quella iniziale può essere corretta solo in circa 80% dei casi. Il caso clinico che proponiamo riguarda un uomo di 71 anni giunto alla nostra osservazione per dolore anale, tenesmo rettale, sanguinamento. L'eplorazione rettale ci ha mostrato una neofromazione dolorosa, di colorito brunastro nel canale anale. La colonscopia e la endoscopia hanno evidenziato la presenza di una grande massa stenotica interessante il canale anale ed il retto con un diametro di circa 90 mm. La biopsia è positiva per melanoma a cellule maligne pigmentate. La TAC ha mostrato un ispessimento della parete rettale e linfonodi nel tessuto adiposo, nel distretto otturatore bilaterale e metastasi polmonari bilaterali. Il dato di laboratorio del Ca 19-9 è nei livelli normali. Il paziente è stato sottoposto a resezione addomino-perineale con dissezione linfonodale. Non ci sono studi dimostranti che la resezione radicale del melanoma primario ano-rettale è associata ad un miglioramento del controllo locale e della sopravvivenza. I pazienti con malattia localizzata dovrebbero essere sottoposti a escissione locale ogniqualvolta ciò sia tecnicamente fattibile. Il ruolo predominante del trattamento chemio radioterapico preoperatorio è quello di ridurre le recidive locoregionale e della cavità pelvica, e per ottenere un più alto tasso di conservazione dell'apparato sfinteriale. Inoltre facilita la rimozione delle potenziali micrometastasi e riduce le metastasi a distanza.

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