Case Report

Primary rectal melanoma - a case report

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Abstrak

Kulit merupakan tempat yang paling sering terjadinya melanoma, diikuti oleh mata, dan yang ketiga adalah daerah anorektal. Melanoma anorektal merupakan kasus yang jarang. Penyakit ini bersifat sangat agresif dengan gejala perdarahan rektum dan perubahan pola defekasi. Pada pemeriksaan proktoskopi kelainan ini terlihat sebagai lesi polipoid tanpa pigmentasi, atau pigmentasi ringan. Histopatologi merupakan diagnosis pasti. Penanganannya memerlukan eksisi radikal dini. Seorang wanita usia 56 tahun datang dengan melanoma pada rektum sepertiga bawah. Kasus ini dilaporkan karena amat jarang dijumpai.

Abstract

The most common site for malignant melanoma is skin, then eye and third is anorectal region. Primary anorectal malignant melanoma is still very uncommon. It is usually very aggressive and presents with altered bowel habit and rectal bleeding. Proctoscopy shows non-pigmented or lightly pigmented polypoid lesion. Histopathology is confirmatory. Early radical excision is mandatory. A 56 year-old female was presented with malignant melanoma of the lower third of rectum. We report this case for its rarity.

Keywords: malignant melanoma, rectum, radical excision

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Primary anorectal melanoma is a very rare and highly malignant disease which constitutes only 0.4-1.6% of all melanomas.[1] This condition was first described by Moore in 1857.[2] Though anorectal region is the third most common site of involvement after skin and eye, yet it accounts only for approximately 0.5% of all colorectal or anal cancers.[3] Nevertheless anorectum is the most common site for development of primary melanoma in the alimentary tract. It has almost equal male to female ratio with an average age of presentation between the fifth and the sixth decade of life.[4] Some studies show female preponderance.[5,6] Majority of patients in the world literatures are Caucasian.[7] Early surgery is the most effective treatment either in the form of wide local excision or abdominoperineal resection with comparable survival,[8] since current chemotherapy and radiotherapy alone have been proven ineffective.

CASE REPORT

A 56 year old female patient presented with burning sensation during defecation and rectal bleeding of 5 months duration with gradually increasing constipation for the last 2 months. General examination was unremarkable except for moderate pallor. Abdominal examination was essentially normal. Inguinal lymph nodes were not palpable. Rectal examination and proctoscopy revealed 4 cm x 3 cm exophytic dark colored mobile firm mass in the right postero-lateral aspect of lower rectum 3 cm above anal verge, with central ulceration and luminal narrowing. The growth bled to touch. A punch biopsy was taken from the growth.

The only abnormality in routine hematological examination was anemia. Colonoscopy showed the

presence of a dark brown exophytic hard mass in lower rectum, rest of colon being normal. Computed tomography of abdomen and pelvis revealed the presence of a large, irregular hypodense mass with heterogeneous enhancement in the rectum. A few enlarged pararectal lymphnodes were present. Bony pelvis was normal. Liver was mildly enlarged with no space occupying lesion. Histopathological examination showed malignant cells with brown pigmentation, suggestive of malignant melanoma.

Patient was counseled about the need for radical excision of tumor and underwent abdominoperineal resection with permanent colostomy. Specimen shows 4 x 4 cm dark colored firm mass with a central ulceration 3 cm above the anal verge (Figure 1). Histopathology suggested malignant melanoma invading perirectal tissue without any lymph node involvement (Figure 2). The immunohistochemistry was positive for S-100, but negative for CA 19-9 and CEA. It confirmed the diagnosis of malignant melanoma. In view of bulky tumor, patient received 6 cycles of chemotherapy. Despite the therapeutic effort, the patient developed multiple liver metastasis after 6 months and died after 9 months.

DISCUSSION

Malignant melanoma is a rare tumor of the rectum with a poor prognosis owing particularly to late presentation. Usual presentation is rectal bleeding, pain, and pruritus. Nearly 5% of patients present with incontinence.[9] Often it is misdiagnosed as rectal polyp or thrombosed piles. Because of these benign symptoms often it is bulky at the time of presentation.[10] One-third of the patients already suffer from dissemination at the time of diagnosis (16-33%).[11] Metastases occur via lymphatic and hematogenous routes. Lymphatic spread to mesenteric nodes is more common than to inguinal nodes. Lungs, liver, and bones are the more frequent sites for distant metastases.[12] Predicted factors for poor prognosis are advanced nature of the disease at the time of diagnosis, ulceration, the rich vascularity of rectal mucosa and high biological aggressiveness of the tumour.[13] Slingluff and Cooper[14] showed 5 year survival rate as less than 10%. Mayo clinic however, reported 5 year survival as 22% and cure in 16% in their study.[6]

Digital rectal examination provides informations regarding size, fixation, and ulceration of the tumor. Endoluminal ultrasound is an established mode of evaluation of the tumor thickness and its nodal status. S-100 protein, Melan A, HMB-45 and MiTF (microphthalmia-transcription-factor) are useful immunohistochemical markers.

In the absence of any metastasis, surgical therapy is mainstay of treatment. Surgical options are abdominoperineal resection (APR) or sphincter preserving wide local excision. Controversy remains about making an option. Former procedure gives better control of local disease, but without clear improvement in survival.[15] Sphincter preserving wide local excision with 2 cm margin and radiotherapy avoids need for stoma, but often is complicated by incontinence or radiation dermatitis. Long term survival is rare as most patients die of disseminated systemic disease regardless of treatment. Some authors do not consider radical surgery as choice of treatment.[13] Survival seems to depend on staging and does not seem to be modified by surgical radicality.[16]

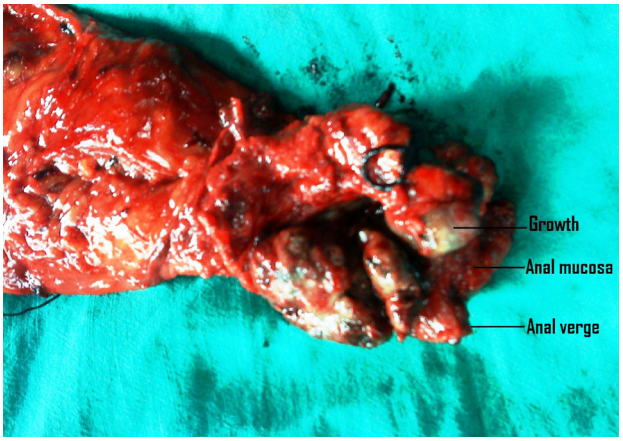

Figure 1. Cut-open lower rectum and anal canal of abdomino-perineal resection specimen showing lower rectal melanoma

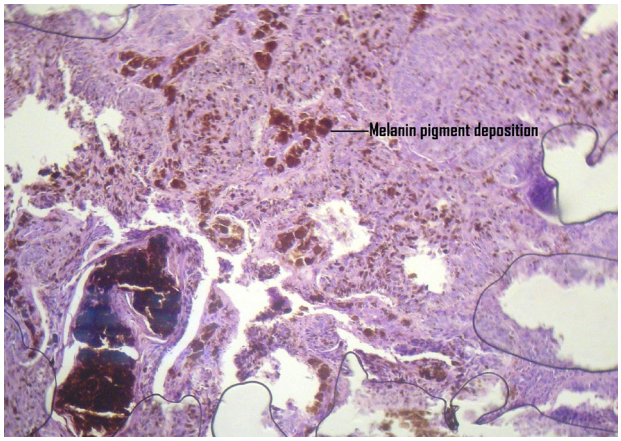

Figure 2. Histopathological examination of the abdomino-perineal resection specimen (HE, x10) showing diffuse melanin pigment deposition

When patients are compared by similar stages, no survival difference has been demonstrated in the two surgical options.[14] Prophylactic lymph node dissection is of no value whereas therapeutic lymph node dissection is indicated only in the presence of inguinal nodes.[17] Severely ill patients, not fit for any surgery, have benefited from intra-tumor beta-interferon injection with systemic dacarbazine.[18] Systemic chemotherapy is indicated for disseminated and lymph node positive diseases. Cisplatin, vinblastin, dacarbazine, INF and interleukin-2 are the chemotherapeutic agents, as used in advanced cutaneous melanoma also.[19]

Tumor thickness seems to be a strong predictive factor for the risk of local recurrences. For anorectal melanoma tumor thickness may also be used as a guideline, i.e. in early disease with a tumor thickness below 1 mm a local sphincter-saving excision with a 1 cm safety margin and in cases of tumor thickness between 1 and 4 mm a local sphincter-saving excision with a safety margin of 2 cm seems to be adequate, whereas tumors with thickness above 4 mm should be treated with APR.[9] Regarding our case, the patient presented with very low bulky rectal melanoma without lymph node involvement and clinically apparent systemic disease. The patient underwent APR.

Regardless of surgical approach anorectal melanoma remains a highly lethal malignancy with overall 5-year survival rate less than 20% according to all reported series.[20]

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