

Case Report of Primary Anorectal Malignant Melanoma- A Rare Aggressive Tumor

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Abstract: Primary Anorectal malignant melanoma (ARMM) is a rare and aggressive tumor that tends to invade locally and metastasize early in the course of the disease. Surgical excision remains the mainstay of therapy. Overall prognosis is poor with a 5 year survival rate of less than 20 %. We report a case of 65 year old female patient who presented with rectal bleeding. She was found to have a mass arising from the anal canal. The patient underwent abdominoperineal resection (APR) with subsequent histopathological examination and immunohistochemistry confirmed the diagnosis of anorectal malignant melanoma.

Keywords: Anorectal, Malignant melanoma, Mucosal

I. Introduction

Primary anorectal malignant melanoma is a rare, aggressive tumor with an unfavorable prognosis and a predilection for early infiltration and distant spread resulting in poor overall survival [1]. It represents approximately 1% of all melanomas and about 0.5-2% of all anorectal malignancies [2]. Typical presentation is in the fifth or sixth decade of life and females are more commonly affected than males [3]. The most common symptoms at presentation are tenesmus, rectal bleeding, changes in bowel habits or a rectal mass [2]. Owing to lack of pathognomonic clinical complaints, it is often misdiagnosed as hemorrhoids or as one of the other benign anorectal conditions [2]. Surgery is the mainstay of treatment, and adjuvant therapies are of limited value. The prognosis of anorectal melanomas remain grim with 5 year survival rate of less than 20% [2,4,5].

II. Case Presentation

A 65 year old female patient presented with history of rectal bleeding and rectal mass of two months duration. Rectal examination revealed a pedunculated growth in the posterior wall of rectum. On colonoscopy, a pigmented, polypoid, ulceroproliferative growth was observed at the anorectal junction (Fig.1). Biopsy was reported as malignant melanoma. CT scan revealed an irregular, intraluminal polypoid mass lesion in the anal canal. Dermatological and ophthalmologic examinations revealed no evidence of cutaneous or ocular primary lesion. The patient underwent abdominoperineal resection and made an uncomplicated recovery. On histopathological examination, a grey black tumor mass measuring 5x5cm was identified 4cm from the anal verge with no involvement of lymph nodes (Fig.2). Microscopy, demonstrated a tumor composed of round cells with round nuclei, coarse chromatin, single prominent nucleolus and scanty cytoplasm. Dense intracytoplasmic melanin deposition was observed (Fig.3). Immunohistochemical confirmation was obtained with strong positivity for HMB 45 (Fig.4) and Melan A.

III. Discussion

Melanomas stem from malignant transformation of melanocytes, which are derived from the neuroectoderm [6]. Although majority of the melanomas are cutaneous in origin, they occasionally arise from extracutaneous tissues that contain melanocytes such as mucosa of the eye, gastro-intestinal tract, respiratory and genito-urinary tracts. Mucosal melanomas account for 1.3-1.4% of all melanomas [6]. First reported by Moore DW in 1857, primary anorectal malignant melanoma is a rare, highly lethal malignancy [2, 7]. It is the third most common location after cutaneous and ocular melanomas [2]. It is the most common primary melanoma of the GIT and accounts for approximately 0.5% of all colorectal and anal cancers. It is more common in females, and the mean age of disease onset is 60 years [1]. The most common symptoms are rectal bleeding, pain, rectal mass and changes in bowel habits [1].

Primary anorectal malignant melanomas are in almost 80% of the cases misdiagnosed as hemorrhoids, polyp, adenocarcinoma or rectal ulcer [2]. Grossly, majority of the lesions appear polypoid, with or without pigmentation and can be ulcerated as well [2]. The tumor is amelanotic in about 30% of the cases [2]. Misdiagnosis is not uncommon, particularly in amelanotic cases with unusual morphologic features that can be mistaken for lymphoma, carcinoma or sarcoma [4,8]. Positivity for immunohistochemical markers such as S100 protein, HMB45 and Melan-A are strongly suggestive of melanoma [1,4]. Four histologic cell types namely epithelioid, spindle cell, lymphoma-like and pleomorphic have been reported by Chute et al [2]. At the time of diagnosis, up to 60% of the patients have metastasis [4]. The 5 year survival rate has been reported to be less

than 20% with dismal prognosis [2,7,9]. Prognostic factors include the stage of the disease at the time of diagnosis and the tumor thickness. Common sites of distant metastasis are the liver and lung [2].

After the histologic diagnosis of anorectal malignant melanoma, a complete staging and search for possible distant metastasis (colonoscopy, CT abdomen and thorax, MRI of the pelvis and brain) as well as ruling out primary sites (skin and retina) are mandatory [7]. Due to its relative rarity, the treatment of anorectal malignant melanoma is controversial [4]. Despite the various modalities available for treatment of anorectal melanoma, surgery has been the cornerstone of therapy [4,7,10]. APR with or without bilateral inguinal lymphadenectomy and wide local excision [WLE] have been used to manage patients with anorectal malignant melanoma [1,3,4,7,10]. However, there is no consensus at this moment on which surgical approach is preferred [3]. APR is considered to reduce the probability of recurrence by controlling the submucosal spread to mesenteric lymph nodes and creating a larger negative resection margin [3,7]. However, available data suggest no significant difference in survival among patients managed with APR and WLE [1,7]. Preoperative tumor thickness may be a valuable tool to plan the surgical approach [2]. APR is the first choice for patients with anorectal melanoma, particularly those with smaller tumors and no evidence of nodal metastasis [1]. APR is associated with high rates of morbidity and colostomy-associated decrease in quality of life. Many authors advocate WLE if negative margins are achievable [7]. Chemotherapy, radiotherapy and immunotherapy should be considered in treatment of anorectal melanoma. However, the results are limited [1]. Despite surgical resection and the emergence of various forms of adjuvant therapy, the overall prognosis of anorectal melanoma remains poor [1,7,10].

IV. Conclusion

Anorectal malignant melanoma is a rare and aggressive neoplasm, which often manifests with advanced disease. In spite of its rarity, anorectal malignant melanoma should be considered in the differential diagnosis of elderly patients presenting with rectal bleeding, tenesmus and change of bowel habits.

References

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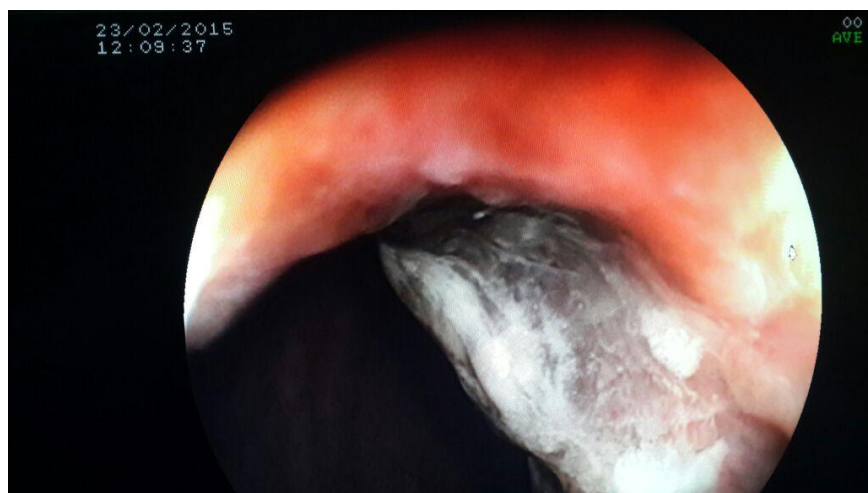


Figure 1: Colonoscopic view of pigmented polypoid growth in the anorectal region



Figure 2: Abdominoperineal resection specimen showing polypoid, grey black growth.

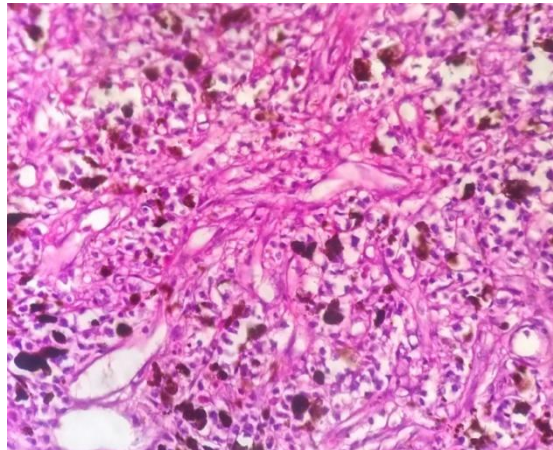


Figure 3: Tumor composed of round cells with prominent nucleoli and dense melanin deposition on Haematoxylin & Eosin stained section at 40X magnification.

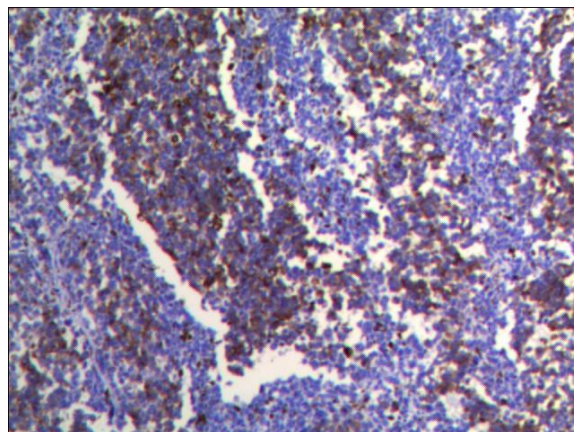


Figure 4: Positivity with immunohistochemical marker HMB 45 at 10X magnification.