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Original Research Paper

General Surgery

PRIMARY ANORECTAL MELANOMA – A RARE CASE REPORT

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ABSTRACT INTRODUCTION: Anorectal melanoma is a rare form of melanoma affecting the anus and/or rectum. It accounts for 0.25-1.25% of all anorectal malignancies and is the third most common site for melanoma, after the skin and eye. It mainly affects elderly age group with no significant gender differences. The most common symptom is per rectal bleeding.

CASE PRESENTATION: A case of 60 year old female presented with blood in stools and difficulty in defecation since a month and on colonoscopy shows polypoidal mass with visual necrosis and haemorrhage, biopsy confirmed anorectal melanoma with HMB 45 positive. On CECT W/A shows pararectal lymph nodes enlarged with no distant metastasis. Stage III A ANM. Later patient underwent APR.

CONCLUSION: Despite the tumour being aggressive in nature and appeals for a less aggressive management while maintaining the quality of life. The prognosis of anal melanoma remains grim and additional research is required to understand better the biology and behaviour of this disease, to assess the better treatment option, meta analysis has to be considered.

KEYWORDS : Anorectal Melanoma, Bleeding, Abdominoperineal Resection.

INTRODUCTION

Anorectal melanoma is a rare form of melanoma affecting the anus and/or rectum. It accounts for 0.25-1.25% of all anorectal malignancies and is the third most common site for melanoma, after the skin and eye.¹Anorectal melanoma affects mainly people aged 50-80 years, with peak incidence in the sixth and seventh decades of life. There are no significant gender differences, whereas the Caucasian race seems to be more frequently affected than African American individuals.¹

The most common symptom prompting presentation of these patients is bleeding^{2,3}. Other symptoms include mass, pain, obstipation, and diarrhea, but pathologic diagnosis after a hemorrhoidectomy is not infrequent.^{2,3,4}

There is currently no standard treatment for anorectal malignant melanoma, largely due to the overall low incidence and lack of evidence in the literature.

The typical therapeutic approach remains surgical resection, it is controversial whether abdominoperineal resection of the anorectum or wide local excision provides the better outcome.

This report present a patient who underwent APR for primary anorectal malignant melanoma and completed 36-months of follow-up.

CASE REPORT

A case of 60 year old female presented with blood in stools and difficulty in defecation since 1 month. The patient presented with no significant weight loss and without any comorbid conditions. She denied any other significant personal and family history.

On digital rectal examination and proctoscopy revealed a polypoidal mass in the anorectal junction at 7 o'clock position. Colonoscopy showed a 200-300 mm polypoidal mass on the posterior wall of the rectum, located within 2cm of the anal verge, and with visual necrosis and hemorrhage (Fig. 1). Biopsies were taken for analysis.



(Fig. 1)

 $Contrast-enhanced \ computed \ tomography \ detected \\ perirectal lymph node and no distant metastasis. Fig-2$

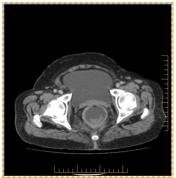


Fig-2

Histopathological examination of the biopsied tissue showedTransanalpolypectomy specimen showedLarge areas of necrosis & haemorrhage Tiny clusters of pleomorphic melanocytes having intracytoplasmic pigmentsIncreased N:C ratio with prominent nucleoli. Thus the diagnosis of anorectal melanoma was made.

Immunohistochemistry analyses were also performed using specific markers to confirm malignant melanoma; the tumor was classified as HMB-45(+).

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Blood tests showed no biochemical abnormalities. Carcinoembryonic antigen and carbohydrate antigen 19-9 levels were also within normal ranges.

The patient underwent APR, fig - 5.

The specimen sent for histopathologic examination Grossing report – Grey white to grey brown soft tissue mass of size $3 \times 2 \times 1$ cm, C/S – Grey white to grey brown.

Microscopic report - Specimen shows Colonic mucosa with focal erosions. Submucosa shows infiltrating neoplasm arranged in sheets and nests, composed of ovoid spindle cells having vesicular nuclei, prominent nucleoli and moderate amount of eosinophilic cytoplasm some of tumor cells contain melanin pigment in their cytoplasm. There is seen brisk mitotic activity in tumorcelss (8-10/10 HPF). Tumorstroma is scant and contain melanophages.

The regional perirectal lymph node shows positive for tumor cells ie N1. Postoperative paraffin pathology of the resected tissues confirmed the diagnosis of stage III A ANM.

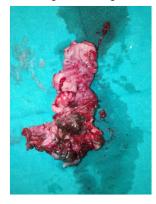


Fig-5

DISCUSSION

AMM is a rare type of melanoma, with a poor prognosis. The incidence of AMM increases with advanced age in both sexes and at all tumor sites. In general, the 5-year survival rate is low, being only6% to 22% reportedly with a median survival period of 12.2 to 22 months. $^{\rm 2.49}$

AMM lacks subjective symptoms in the early stage. The most common symptom is bleeding, with 53% to 89% of patients reporting this as the predominant complaint. The other symptoms include altered bowel habits, constipation, decreased stool caliber, unintentional weight loss, and palpable inguinal mass. Unfortunately, quite a few patients are diagnosed with AMM when they have already developed distant or regional metastasis.¹⁰Weinstock reported that 41% of anorectalmelanoma cases had regional spread and 22% had distant metastasis, while 37% had confined disease.¹¹The major sites of distant metastases are lung, liver, brain, bone, and breast. $^{\rm 12\cdot14}$

Our patient had no distant metastasis. Histopathologically, AMM show variability regarding the size and type of cells. It can be misdiagnosed as malignant lymphoma, small round cell sarcoma, spindle cellsarcoma,gastrointestinal stromal tumour, and epidermoid carcinoma. Thus, immunohistochemical analysis plays a pivotal role in the diagnosis of AMM. Anti-S-100 protein, human melanoma black (HMB-45), Vimentin, and Melan A antibody are the melanocyte specific stains used for diagnosis of malignant melanoma. Our patient showed positive for HMB-45.

The treatment of anorectal melanoma is controversial. While the typical therapeutic approach remains surgical resection, there is no consensus on which surgical approach -WLE or APR - is preferred. APR is regarded as the standard surgery for treatment of AMM because it can control lymphatic spread and obtain a larger negative margin for local control.^{15,}

The patient in this study underwent APR without adjuvant therapy. Till date, she has been attending regularfollow-up every 3 to 4 months and no complications or recurrences in this current condition have presented.

CONCLUSION

Despite the tumour being aggressive in nature and appeals for a less aggressive management while maintaining the quality of life. The prognosis of anal melanoma remains grim and additional research is required to understand better the biology and behaviour of this disease, to assess the better treatment option, meta analysis has to be considered.

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