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Primary Ano-rectal Melanoma- A Rare but lethal entity

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ABSTRACT:

Background: Primary ano-rectal malignant melanoma (ARMM) is rare but aggressive tumor**.** After skin and retina though this is the third most common site of melanoma. It accounts only for approximately 0.5% of all colorectal or anal cancers. Symptoms include passing of blood with stools, anal pain, and changes in bowel habits or rectal mass.

Material and methods: The study was carried in the department of pathology sher-i-Kashmir institute of medical sciences soura Srinagar. A total of five cases that underwent surgery for anorectal mass and diagnosed as primary anorectal melanoma were included in the study. Retrospective records of 8 years (January 2010 to December 2017) were examined.

Results: 5 cases of ano-rectal malignant melanoma were identified. Out of 5 patients, 60% were females and 40% were male patients. Age of the patients ranged from 60-75 yrs. The commonest presenting symptom was blood with stools 60% of the patients. Most of the patients at the time of diagnosis had Stage III disease (60%) with nodal metastasis in all the patients. The commonest histological variant seen was epitheloid variant of malignant melanoma (3 cases).

Conclusion: Elderly patients presenting with non-specific symptoms like melena, mass prolapse, and abnormal bowel habits: malignant melanoma should always be kept as one of the differential diagnosis.

Key words: Melanoma, APR, Melena, IHC, WLE

Introduction:

Primary anorectal malignant melanoma (ARMM) is rare but aggressive tumor (1). After skin and retina though this is the third most common site of melanoma (2) but accounts only for approximately 0.5% of all colorectal or anal cancers (3) With an incidence of 0.5–4% of all the malignancies (4). Typical presentation being fifth to sixth decade of life, commonly affecting women (5). Symptoms that are common to other tumors of anorectal region such as passing of blood with stools, anal pain, changes in bowel habits or rectal mass can be seen in melanoma also (6). Treatment options including wide local excision (WLE), abdomino-perineal resection (APR), chemotherapy and radiotherapy, but ARMM is frequently radiotherapy resistant and shows a poor response to chemotherapy (7). As anal melanoma is rare, only small case series have been reported in the literature, making it difficult to draw conclusions about optimal treatment and outcome (8).

Material and methods:

The study was carried in the department of pathology sher-i-Kashmir institute of medical sciences soura Srinagar. Retrospective records of 8 years (January 2010 to December 2017) were examined. A total of five cases who underwent surgery for anorectal mass and diagnosed as primary anorectal melanoma were included in the

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study. Relevant blocks were taken and sections re-examined by two separate pathologists to reduce the bias and further subjected to immunohistochemistry (IHC) for confirmation of the diagnosis.

Results:

5 cases of anorectal malignant melanoma were identified from January 2010 to December 2017. Out of 5 patients, 3 (60%) were females and 2 (40%) were male patients. Age of the patients ranged from 60-75 yrs. The commonest presenting symptom was bleeding per rectum (blood with stools) in 3 (60%) patients, 1 (20%) patient presented with history of constipation and 1 (20%) patient presented with worsening fecal incontinence over several months associated with sharp rectal pain (Table-1). Other associated symptoms included change in bowel habits, weight loss, tenesmus and anorexia. Proctoscopic examination showed that the tumor was visible in 3 cases (60%) and palpable in almost all cases. The pathological characteristics of patients with ano-rectal melanoma are shown in Table -2.

Table 1: Distribution of cases according to age, gender, size, clinical presentation and surgical procedure.

Age (yrs)	Gender	Clinical presentation	Tumor size (cm)	Surgical procedure
60	Female	Bleeding per rectum	2x1x1	WLE
63	Male	Bleeding per rectum	4x3x2	APR
65	Female	Bleeding per rectum	5x5x4	APR
72	Female	Fecal incontinence and rectal pain	5x3x2	APR
75	Male	constipation	2x1x0.5	WLE

Table 2: Pathological characteristics of patients diagnosed with anorectal melanoma.

Features	Number of patients	Percentage (%)
LVI		
Present	4	80%
Absent	1	20%
PNI		
Present	2	40%
Absent	3	60%
T category		
T1	1	20%
T2	1	20%
T3	3	60%
T4	0	-
N category		
N0	0	-
N1	2	40%
N2	3	60%

TNM Stage Stage I Stage II Stage III Stage IV	 1 1 3 0	 20% 20% 60% -
Mucosal melanoma stage Stage I Stage II Stage III	 1 1 3	 20% 20% 60%

**LVI- lympho-vascular emboli, PNI- peri-neural infiltration*

Most of the patients at the time of diagnosis had Stage III disease (60%) with nodal metastasis in all the patients. 4 patients had died within first ten months after diagnosis and treatment. One patient was on follow up undergoing postoperative chemotherapy. The commonest histological variant seen was epitheloid variant (3 cases) (fig-1 and fig-3) and spindle cell variant (2cases). Melanin pigment (fig-2) was found in 4 cases and was absent in 1 case. IHC was done in all the 5 cases which showed diffuse positivity for HMB 45 and S-100.

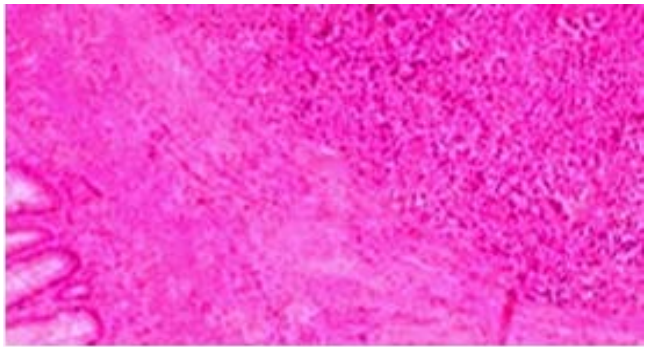

Fig. 1: showing low magnification H&E section revealing tumor cells in sheets with desmoplastic response.

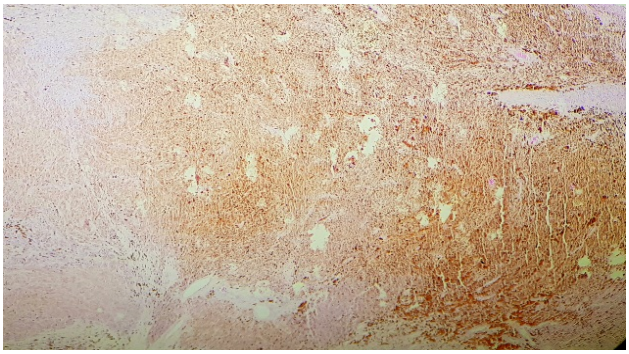

Fig. 2: Extensive melanin pigment within the tumor.

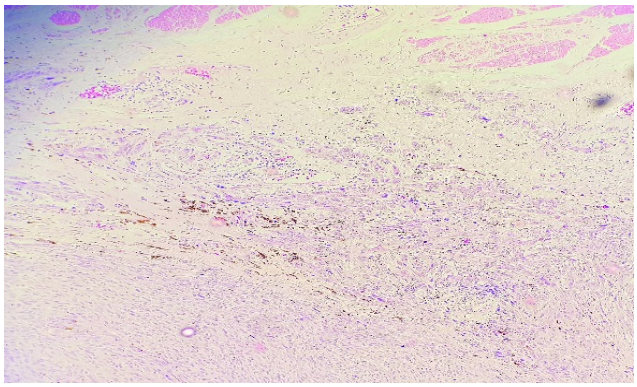

Fig. 3: High magnification showing epitheloid tumor cells having abundant cytoplasm, prominent eosinophilic macronucleoli and predominantly intracellular melanin pigment (H&E).

Discussion:

Anorectal melanomas are aggressive but rare tumors. Compared to that of cutaneous melanomas, anorectal melanoma has lowest percent of five years survival with overall survival time of 10-19 months after diagnosis and survival rate being only 18%. However, women with ARMM displayed a significantly greater longevity than men. (9). Due to absence of specific symptoms and comparing with other anorectal disease, anorectal melanoma is often diagnosed accidentally (10). Anorectal melanoma is commonly seen among individuals in their 60s and often found in women. Patients present with non-specific symptoms like melena, sensation of a mass in the anal region, mass prolapse, and abnormal bowel habits (11). It can be misdiagnosed as rectal polyp or thrombosed piles. Because of these non-specific symptoms it is

bulky at the time of presentation (12). Chute et al evaluated 17 cases of primary ARM with special reference to histopathology and IHC. In their study they found morphological subtypes of ano-rectal melanoma (ARM) were: epithelioid (12 cases), spindle-cell (7 cases), lymphoma-like (10 cases) and pleomorphic (6 cases). Melanin pigmentation was present in nine cases; junctional change was present in six cases and mitotic rate was three or more per high-power microscopy field in eight cases (13). S-100 protein, Melan A, HMB-45 and MiTF (microphthalmia-transcription-factor) are useful immuno-histochemical markers (14). Most studies show a higher rate of local control for patients undergoing APR, but typically without any survival benefit when compared to WLE. The advantages of APR are its ability to control lymphatic spread and to create wider excision margins resulting in a lower local recurrence rate. WLE offers seemingly equivalent symptom control and the opportunity for cure with significantly less morbidity and avoidance of a permanent colostomy, however, there is a higher rate of local recurrence associated with WLE (15). Optimal treatment is still controversial. Patients undergoing WLE or APR, do not demonstrate any survival advantage with either approach (16).

Conclusion:

Elderly patients presenting with non-specific symptoms like melena, sensation of a mass in the anal region, mass prolapse, and abnormal bowel habits: melanoma should be kept in mind as one of the differential diagnosis.

Conflict of interest: Nil.

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