ACADEMICIA
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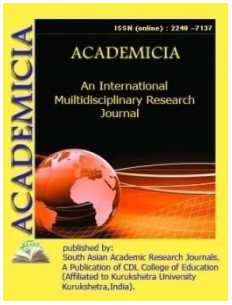

DOI: 10.5958/2249-7137.2021.01893.0

PRIMARY VAGINAL MALIGNANT MELANOMA: A RARE ENTITY

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ABSTRACT

Primary tumour of vagina is a rare neoplasm and known to constitute approximately (1-2%) of all gynaecological malignancies. Among vaginal tumours, malignant melanoma of vagina is known to arise from the region of atypical melanocytic hyperplasia. Vaginal Melanoma is extremely rare neoplasm and known to occur in approximately 4% of all primary vaginal tumours. Incidence of vaginal neoplasm is 0.26% per million every year globally. In term of prognosis and overall survival, Primary vaginal malignant melanoma is known to rare a very poor overall survival. Melanoma vagina is known to have early haematogenous spread and distant spread. Prognosis is very poor for vaginal melanoma due to aggressive nature of disease. Risk of distal metastatic 0.5 year overall survival 05% to 30% and chances of distant metastatic (66-100%). Thus, overall survival of primary malignant melanoma vagina is poor. We report here an unusual case of female diagnosed with primary vaginal malignant melanoma.

KEYWORDS: *Malignant Melanoma, Female, Genital Tract, Primary Vaginal Malignant Melanoma, Rare*.

INTRODUCTION

Vagina is around (3-4) inches long and known to extends from cervix to vestibule. It lies in between urethra and rectum i.e dorsal to urethra and ventral to rectum. Embryologically, vagina lies dual origin i.e upper 1/3red derived from uterine canal and lower 2/3rd derived from urogential sinus. Primary vaginal tumours are rare condition and known to constitute nearly 1-2% of all gynaecologic malignancies. Among all primary vaginal tumours, vaginal melanoma is known to constitute 4% of all primary vaginal tumours. Incidence of primary vaginal tumour melanoma is 0.26 per million per year globally. Incidence increase as the age of females increase incidence rate were high for African-American females compared to Asian females. Vaginal melanoma is highly malignant tumour and is known to have early haematogenous spread. Vaginal melanoma usually at the time of diagnosis present as distant metastasis. Most common symptom of vaginal melanoma is bleeding per vaginum. Most common factor for development of primary vaginal melanoma is presence of pre-invasive condition. Other risk factor includes infection HPV 16 and HPV 18 is known to affect and predispose to cancerous condition. Vaginal melanoma usually appears as blackish, dark mass or ulceration.

Case report: A 55 years old female presented in Radiation Oncology OPD at our institute with chief complaint of bleeding per vaginum and whitish foul smelling discharge per vaginum for 2 months. Patient was apparently well when patient presented with bleeding per vaginum. It was insidious in onset, with fresh blood and clots. Patient also gave history of whitish discharge per vaginum insides in onset and discharge was associated with foul smell.

Past history: Patient had history of (TAH + BSO)-15 years back. Patient had history of hypertension for which she was on oral anti-hypertension drugs from part of 10 years. She has no history of DM, Asthma.

Operation note: Patient underwent evasion of lump from left labia major at same private hospital. After surgery patient was referred to Radiation Oncology OPD at our institute.

CEMRI-abdomen + pelvis – CEMRI showed

1) Hepatomegaly with fatty liver.
2) Slightly small sized left kidney compared to right side.
3) Mild B/L renal cortical irregularity or be due to scaring

PET CT:

-Non-hyper metabolic to hyper metabolic soft tissue thickening in the left inguinal region with evidence of ulceration and seroma formation-suggestion of postsurgical inflammatory sequelae.

-Mild hyper metabolism in the vagina, predominantly towards the left side of the middle.

Fig 1: ***Fig 2:***

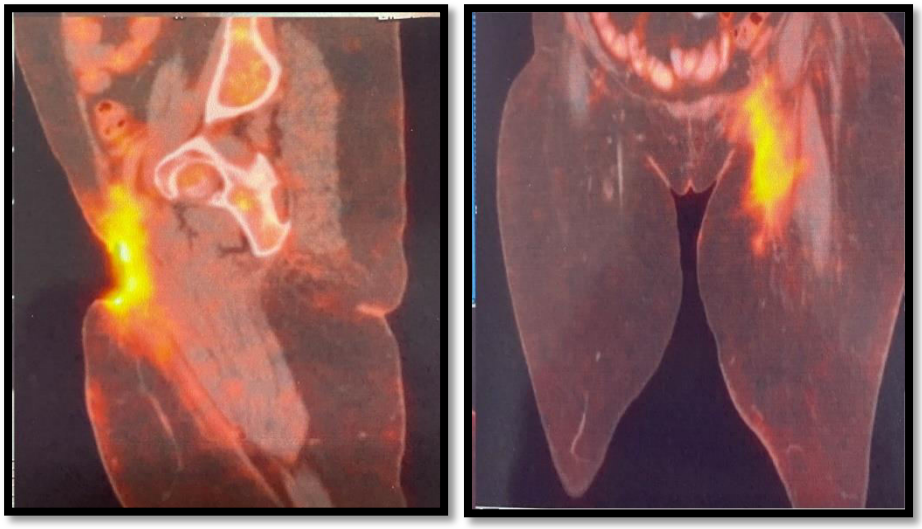


Fig 3:

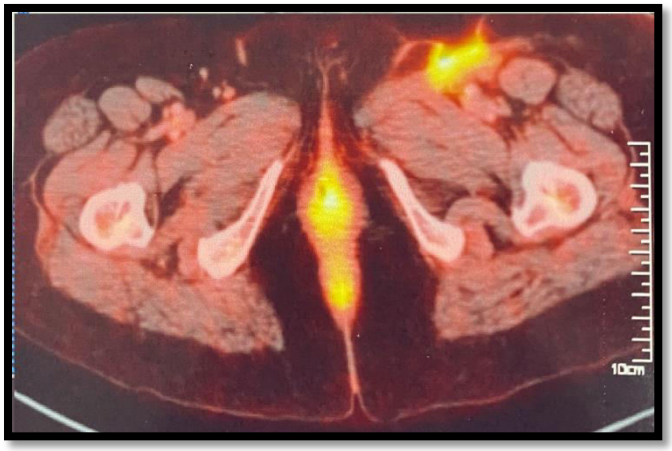


Fig 1, Fig 2 & Fig 3: Shows hypermetabolic lesion at primary site (Vagina) and hypermetabolic activity at loco-regional site.

Histopathological Examination:

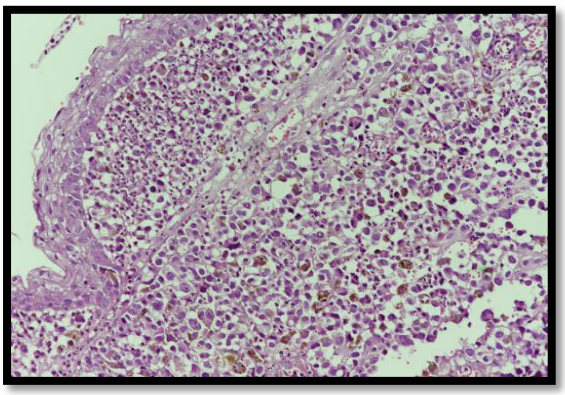


Fig 4: 200 X tumour arranged in lobules

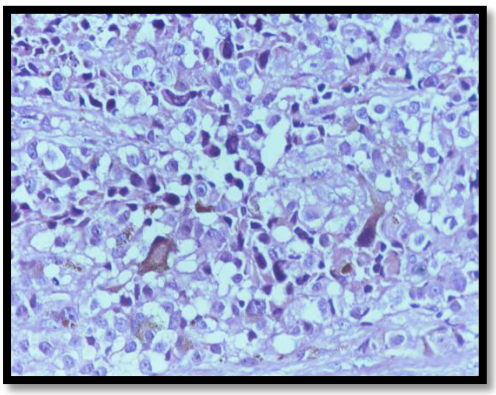


Fig 5: 400 X and markedly anaplastic

Showed multiple fragments which are focally lined by stratified squamous epithelium. Sub epithelium shows as tumour arranged in lobules. Tumour cell are markedly anaplastic having vesicular nuclear chromatin, prominent eosinophillic nucleoli and moderate amount of vacuolated cytoplasm with abundant intracellular and extra cellular melanin.

Immunohistochemistry:

Fig 6 & 7 Tumour cells are positive for (HMB-45 AND S-100 Protein)

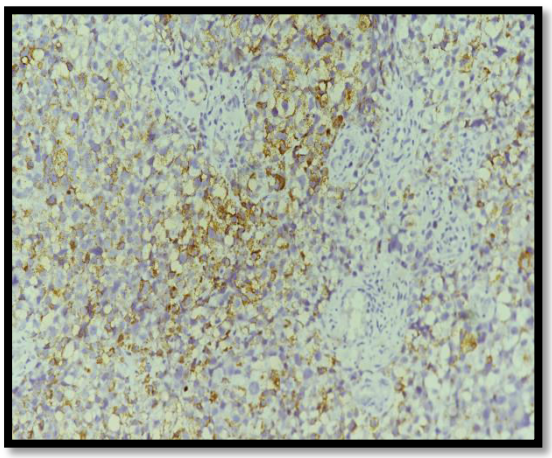
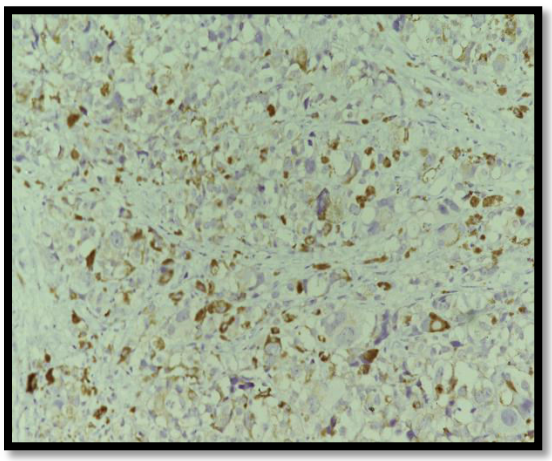


Fig: 6



Figs: 7

Treatment:

Primary vaginal malignant melanoma is extremely rare neoplasm. So, very few studies are reported in literature for management of vaginal melanoma. Treatment approach include-surgery, chemotherapy and radiotherapy either singly or combined modality.

Primary treatment of choice for vaginal melanoma is surgery. Surgery is preferred over radiotherapy or malignant melanoma is radio resistant tumour. So, surgery is preferred option. Surgery includes radical surgery and wide local excision. There is no role for lymadenopathy in vaginal cancer. As, lymadenopathy ultimately leads to lymphedma which contribute to patient's

morbidity. Whenever radiation is added as an adjuvant treatment modality, the dose of Radiation is kept ranging to 50 gray to 75 gray. As melanoma is known to show systematic spread very early. So, adjuvant chemotherapy is known add some benefits in local control. Thus, Adjuvant (RT and CT) is known to shows some degree of improvement in local control. Many other targeted are also there for vaginal melanoma. These targeted therapies include.

a) Ipilimumab (antibody against cytotovic T.lyphocyte-associated protein 4) (CTLA-4)

b) Nivolumab (PD-1 inhibitor)

c) Vemurafenib (BRAF-inhibitor)

d) Dabrafenib (BRAF-inhibitor)

-By using multiple combined modality therapy, still overall survival is very poor for vaginal melanoma. 5year overall survival rate is 5-30%.

CONCLUSION:

As vaginal melanoma is very rare (extremely rare) tumour. Overall incidence is first 0.26 per million per year globally. Cancer is most seen in females of older age compared to younger. Females of Africa and America are commonly affected compared to Asia. As, vaginal melanoma is highly malignant tumour and known to show distant metastasis at early age. So, Aggressive treatment approach is very required. Treatment approach includes surgery, chemotherapy and adjuvant radiotherapy. Even after continued modality and aggressive therapy approach, overall 5 year survival is very poor 5-30%. The risk of distant metastasis is around (66-100%). As, primary vaginal malignant melanoma is extremely rare neoplasm and overall risk of distal metastasis is high and prognosis is very poor. There is no clear guideline for treatment of vaginal melanoma. So, further reports of such rare cases 1% in literature us of paramount important to guide the disease risk factor, prognosis and treatment.