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Primary Vaginal Ewing's Sarcoma or Primitive Neuroectodermal Tumor with Liver and Lung Metastasis in a 45-Year Old Woman

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1. Introduction

Ewing'sSarcoma/PNET of the female genital tract is very unusual, but has been reported to involve the ovary, uterine corpus, uterine cervix, and vulva. To our knowledge, only 10-12 cases of primary vaginal Ewing'sSarcoma/PNET have been previously reported in the literature and none of them had any evidence of metastasis when reported. Here, we present a rare case of primary vaginal Ewing's Sarcoma/PNET with liver, breast and lung metastasis.

2. Case Report

We present a case of a 45 year old woman, gravida 2, para 2, with who presented with the complaints of whitish, foul smelling vaginal discharge and swelling at vulva since 2 months and itching at the local site since 1 month. Per vaginal & per speculum examination of vagina showed 6X6 cm submucosal growth over left sided vulva, disease extended from 10'O clock to 5'O clock position of middle & lower vagina but cervix was free.

Figure 1: Growth at left side of vagina extending up to vulva

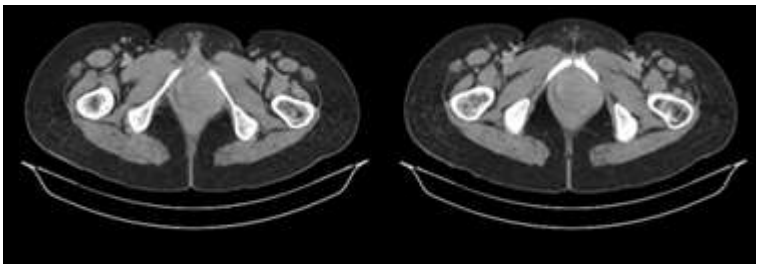

Figure 2A & 2B: Lesion with internal necrotic area involving vagina more on left side and extending upto labial fold

Rectal examination revealed b/l paravaginal tissue medially involved but the rectal mucosa was free. Routine haemogram, liver and renal function tests were within normal limits. Chest radiograph revealed no abnormality and Contrast-enhanced Computed Tomography (CECT) of Thorax revealed few calcified nodes in right hilar region and sub-carinal region with bilateral lung metastasis and liver metastasis. CECT Abdomen revealed liver metastasis. CECT Pelvis showed a 57X47X120 mm lesion with internal necrotic area involving vagina more on left side and extending upto labial fold, both is chiorectal fossa and infiltrating the proximal part of the leftobturator internus, loss of fat plane with rectum and anal canal; 40X38mm fibroid involving fundus of uterus;bilateral adnexa were normal with no ascites or lymphadenopathy. Bone scan was normal. Punch biopsy of the vaginal mass was then performed which showed poorly differentiated Adenocarcinoma with probable Neuroendocrine

differentiation. Immunohistochemistry was done with a panel of antibodies, which revealed Ewing's Sarcoma. Following our diagnosis of Primary Ewing's sarcoma or PNET of the vagina, our patient was subjected to combination chemotherapy for 35 days 1 cycle VACA (Vincristine, Actinomycin-D, Cyclophosphamide and Doxorubicin). During chemotherapy disease was progressive, so the patient was sent for palliative radiotherapy and was treated with 30Gy/15# (@200cGy/#) by AP/PA portals, during which our patient was found to have clinically progressive disease. Following this, she was on palliative chemotherapy, single agent (Adriamycin).

Table 10: Cases of primary vaginal Ewing's sarcoma/PNET reported earlier

Study	Age	T-size	IHC profile	Treatment	Follow up(months)	Outcome
Liao et al	30	5	VIM+, MIC2+,FLI+,Synaptopysin+,NSE+,S-100+	TAH+BSO+CT	36	FOD
Farley et al	35	4	MIC+	CT+EBRT+ICBT	48	FOD
Vang et al	35	3	VIM+,MIC2+	WE+CT+RT	19	FOD
Gaona-luviano et al	34	4	MIC2+	WE+CT+RT+ICBT	20	FOD
Rekhi ET AL	17	10	VIM=,MIC2=,FL1=,BCL2=	CT+EBRT	FU	
Al-Taimini et al	47	ND	ND	ND	ND	ND
Yip et al	27	6	MIC2+	WE+RT	18	FOD
Pang et al	54	4	MIC2+	EBRT+ICBT	18	DOD
Petkovic et al	45	9	MIC2+	CT+EBRT+ICBT	18	AWD
McCluggage et al	40	8	VIM+,MIC2+,FL1+	ND	ND	
Our case	45	11	VIM+, MIB1+(>50%), CD99+	CT+EBRT+CT	FU	

PNET = primitive neuroectodermal tumour; T-size = tumour size in largest dimension; IHC=immunohistochemistry; VIM = vimentin; + = positive; – = negative; WE = Wide excision; CT = chemotherapy; EBRT = external beam radiotherapy; ICBT = intracavitary brachytherapy; TAH+BSO = total abdominal hysterectomy + bilateral salpingoophorectomy; MIC2 = Microneme protein 2; FLI1 = FOD = free of disease; AWD = alive with disease; DOD = died of disease; FU = follow-up; ND=not described

3. Result

Our patient was regular in treatment.

4. Discussion

Ewing's sarcoma has a potential for haematogenous metastasis and the most common sites of metastasis include lungs, bones and bone marrow. About 25% of patients have metastatic disease at presentation, patients with isolated lung metastasis have better prognosis than those with extra-pulmonary disease. The chemotherapy regimen and initial treatment for patients with metastatic disease is the same as that for localized disease. At the time of local therapy, all sites of the disease must be re-evaluated. If tumor shows progression or there is persistence of widespread disease, there is little hope for cure and hence, such patients should be treated with palliative intent. For patients responding well, at this stage, local therapy in the form of surgery and/or radiation is recommended to the primary site as well as all metastatic sites. Management of vaginal Ewing's sarcoma is controversial, due to rarity of its presentation.

5. Conclusion

Our case report describes a rare site of primary vaginal Ewing's sarcoma/PNET in the 45 year old patient. It reinforces the value of IHC, emphasizing the utility of immunohistochemical staining in establishing the diagnosis of tumours at unusual sites. Further the case also highlights the utility of induction chemotherapy followed by radiation treatment and subsequent palliative chemotherapy as a treatment modality.

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