

Primary vaginal Ewing's sarcoma or primitive neuroectodermal tumor with liver, breast and lung metastasis in a 45-year old woman

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Introduction

Ewing sarcoma/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 sarcoma/PNET have previously been reported in the English literature and all of them had no evidence of metastasis when reported. Here, we present a rare case of primary vaginal Ewing sarcoma/PNET with liver, breast and lung metastasis (Figure 1A & 1B).

Case report

We present the case of a 45-year old woman, gravida 2, para 2, with complain of whitish, foul smelling vaginal discharge & swelling at vulva since 2 months, itching at local site since 1 month. Per vaginal & per speculum examination of vagina showed 6*6 submucosal growth at left side vulva, disease involving 10'O'clock to 5'O'clock position of middle & lower vagina, cervix free.

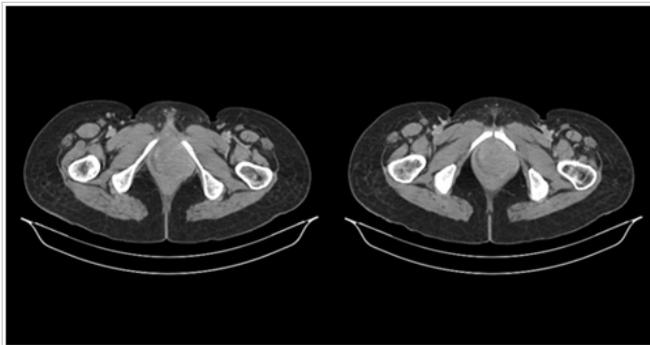


Figure 1A & 1B lesion with internal necrotic area involving vagina more on left side extending upto labial fold.

Rectal examination- B/L paravaginal medially involved rectal mucosa free. Routine haemogram, liver and renal functions are within normal. Chest radiograph revealed no abnormality and contrast-enhanced computed tomography (CECT) thorax revealed few calcified nodes in right hilar region and sub-carinal region sized nodule with bilateral lung metastasis and liver metastasis, heart, great vessels, bilateral bronchi were normal. CECT abdomen revealed liver metastasis both right and left kidneys normal other organs and biliary tree were normal; no abdominal lymphadenopathy was evident. Pelvic CECT scan showed a 57*47*120mm lesion with internal necrotic area involving vagina more on left side extending upto labial fold, both ischiorectal fossa, infiltrates proximal part of

left obturator internus, loss of fat plane with rectum and anal canal, 40*38mm fibroid involving fundus of uterus, Bilateral adnexa were normal with no ascites or lymphadenopathy. Bone scan is 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, during chemotherapy disease was progressive, then patient was sent for palliative radiotherapy 30Gy/15# (200CGy/#) by AP/PA portal, during which our patient was found to be clinically progressive disease. Following this she was on palliative chemotherapy, single agent (Adriamycin) (Table 1).

Result

Our patient is regular in treatment.

Discussion

Ewing's sarcoma has a potential for haematogenous metastasis and the most common sites of metastases 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

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cure 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 sarcoma is controversial, due to rarity of its presentation.^{11,12}

Table 1 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., ¹	30	5	VIM+, MIC2+,FLI+,Synaptophysin+,NSE+,S-100+	TAH+BSO+CT	36	FOD
Farley et al., ²	35	4	MIC+	CT+EBRT+ICBT	48	FOD
Vang et al., ³	35	3	VIM+,MIC2+	WE+CT+RT	19	FOD
Gaona-luviano et al., ⁴	34	4	MIC2+	WE+CT+RT+ICBT	20	FOD
Rekhi et al., ⁵	17	10	VIM=,MIC2=,FLI=,BCL2=	CT+EBRT	FU	
Al-Taimini et al., ⁶	47	ND	ND	ND	ND	ND
Yip et al., ⁷	27	6	MIC2+	WE+RT	18	FOD
Pang et al., ⁸	54	4	MIC2+	EBRT+ICBT	18	DOD
Petkovic et al., ⁹	45	9	MIC2+	CT+EBRT+ICBT	18	AWD
McCluggage et al., ¹⁰	40	8	VIM+,MIC2+,FLI+	ND	ND	
Our case	45	11	VIM+, MIBI+(>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; FOD, free of disease;AWD, alive with disease; DOD, died of disease; FU, follow-up; ND, not described

Conclusion

Our case report describes a rare site of primary vaginal Ewing's sarcoma/PNET in the 45year 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.

Acknowledgments

None.

Conflicts of interest

Authors declares there are no conflicts of interest.

References

- Liao X, Xin X, Lü X. Primary Ewing's sarcoma-primitive neuroectodermal tumour of the vagina. *Gynecol Oncol*. 2004;92:684–688.
- Farley J, O'Boyle JD, Heaton J, et al. Extrasosseous Ewing sarcoma of the vagina. *Obstet Gynecol*. 2000;96(5):832–834.
- Petkovic M, Zamolo G, Muhvic D, et al. The first report of extrasosseous Ewing's sarcoma in the rectovaginal septum. *Tumori*. 2002;88(4):345–346.
- Vang R, Taubenberger JK, Mannion CM, et al. Primary vulval and vaginal extrasosseous Ewing's sarcoma/peripheral neuroectodermal tumour: diagnostic confirmation with CD99 immunostaining and reverse transcriptase polymerase chain reaction. *Int J Gynecol Pathol*. 2000;19(2):103–109.
- Rekhi B, Qureshi S, Basak R, et al. Primary vaginal Ewing's sarcoma or primitive neuroectodermal tumour in a 17-year-old woman: a case report. *J Med Case Rep*. 2010;4:88.
- Al-Tamimi H, Al-Hadi AA, Al-Khater AH, et al. Extraskelatal neuroectodermal tumour of the vagina: a single case report and review. *Arch Gyneco Obstet*. 2009;280(3):465–468.
- Yip CM, Hsu SS, Chang NJ, et al. Primary vaginal extrasosseous Ewing sarcoma/primitive neuroectodermal tumour with cranial metastasis. *J Chin Med Assoc*. 2009;72(6):332–335.
- Pang X, Chen P, Wen F, et al. Primary Ewing's sarcoma/primitive neuroectodermal tumour of the vagina in a 54-year-old woman: a case report. *Arch Gynecol Obstet*. 2012;285:1031–1033.
- Gaona-Luviano P, Unda-Franco E, González-Jara L, et al. Primitive neuroectodermal tumour of the vagina. *Gynecol Oncol*. 2003;91(2):456–458.
- McCluggage WG, Sumathi VP, Nucci MR, et al. Ewing family of tumours involving the vulva and vagina: report of a series of four cases. *J Clin Pathology*. 2007;60(6):674–680.
- Ranjini Kudva, Lakshmi Rao, Mohammed Musheb. Peripheral Primitive Neuroectodermal Tumor (PNET) of the Paravaginal Tissue Ranjini Kudva, Lakshmi Rao, Mohammed Musheb Dept of Pathology, Kasturba Medical College, Manipal, Manipal University, India. *International Journal of Scientific and Research Publications*. 2013;3(3):1–3.
- Das P, Gunaseelan K, Basu D, et al. Primary vaginal ewing's sarcoma/primitive neuroectodermal tumour: diagnostic and treatment challenges. *J Clin Sci Res*. 2014;3:145–149.