

PRIMARY VAGINAL EWING SARCOMA: A DIAGNOSTIC AND THERAPEUTIC CHALLENGE

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Objective:

This case report details the treatment of a young woman with an unusual tumour that necessitated close coordination between geneticist, pathologist, radiation oncologist, and surgeon. A primary neuro-ectodermal tumour, vaginal Ewing's sarcoma develops in soft tissue or bone. It is extremely uncommon and accounts for 1- 2% of all gynaecological malignancies.

Case report:

A 30-year-old nulliparous lady complained of vaginal discharge for two months. A 6 cm vaginal lump was discovered during imaging and clinical evaluation.

Diagnosis:

According to immunohistochemistry, the vaginal mass biopsy revealed poorly differentiated carcinoma with positive CD-99, Cyclin-D1, and FLI-1 proteins. The patient was submitted for genetic screening, the FISH assay (fluorescence in situ hybridization), and gene testing for EWSR1 (EWS RNA binding protein 1). Vaginal Ewing's sarcoma was confirmed by genetic testing (EWS RNA binding protein 1). This confirmed the diagnosis of germ line mutation-associated vaginal Ewing's sarcoma with chromosomal 22q12 translocation.

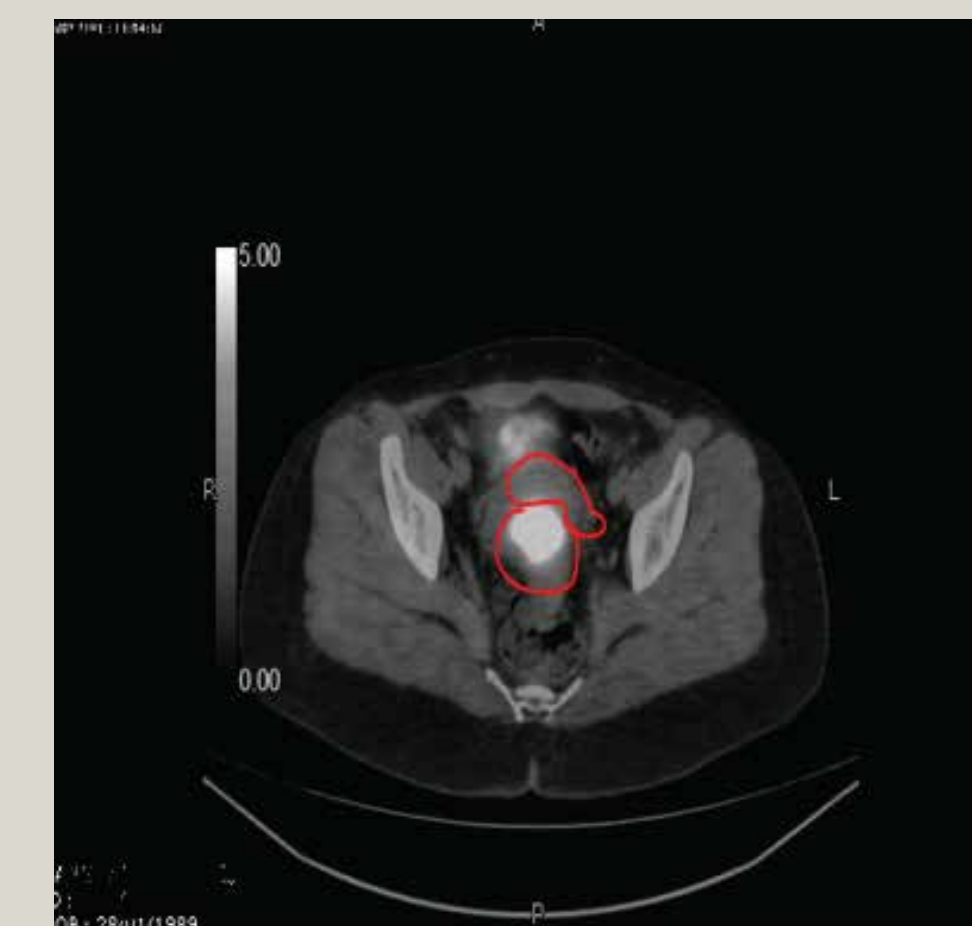
Treatment:

Multidisciplinary teamwork was required for her management and following the freezing of embryos during the IVF cycle to preserve fertility, Her first line neoadjuvant chemotherapy started with VAC/IE regimen for 6 cycles (Vincristine, Adriamycin, Cyclophosphamide, Ifosfamide, Etoposide) followed by radiation therapy. During her follow-up visit while on neoadjuvant treatment regression of tumor seen on clinical examination as well radiological imaging with MRI and PET/CT scan, so she underwent conservative surgery with MDT review. Her wide local excision of vaginal mass with free margins was done which showed foci of residual Ewing's Sarcoma

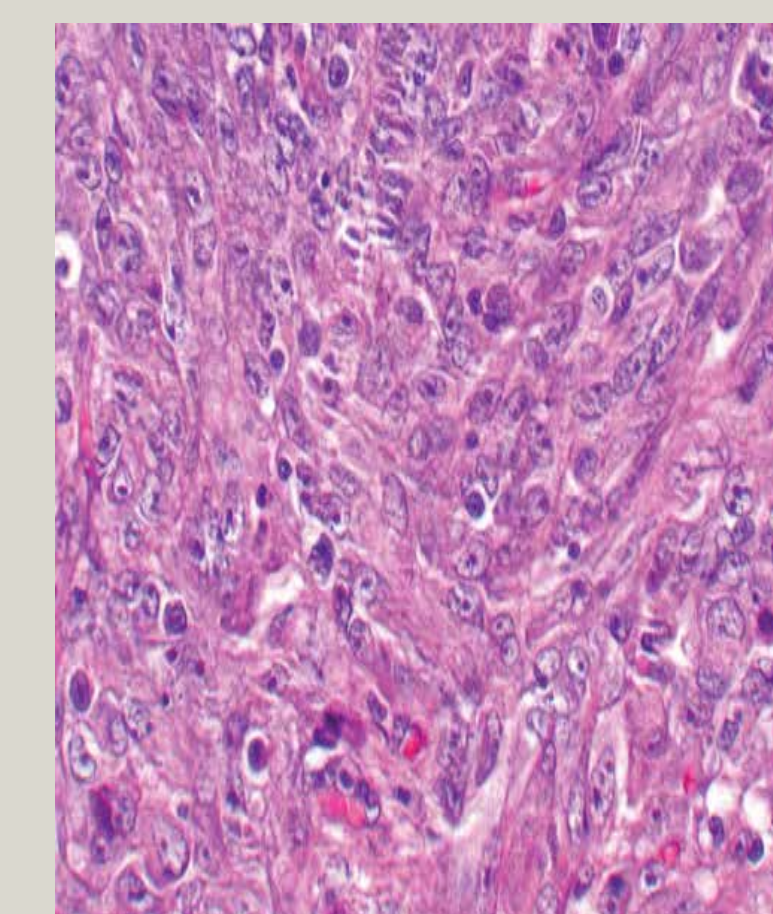
Figure: 1 MRI pelvis with contrast.



Figure: 2 PET/CT scan with contrast.



- Histopathological Diagnosis:
- Vaginal biopsy result:
- Poorly differentiated carcinoma / Ewings sarcoma
- CD99 Positive
- Cyclin-D1 Positive
- FLI-1 Positive
- Cytokeratin AE1/AE3 Dot positive
- **Friend leukemia integration 1 transcription factor**



Treatment after disease progression:

- Again During radiation fractionated treatment progression of disease seen on local vaginal examination with left sided nodule at previous tumor site with 2cm deep margins. Her case was again discussed in tumor board meeting and after literature search it was decided to start second line chemotherapy with VIT regimen for 6cycles (vincristine, irinotecan, temozolomide) with pazopanib also included in this regimen.. She received 3 cycles followed by definitive surgery after again detail counselling of couple. She underwent cystoscopy with left DJ ureteric stenting, type C1 redical hysterectomy with bilateral salpingo oophorectomy, enbloc low anterior resection of rectum with mass, sigmoid end colostomy and partial vaginectomy
- Second line chemotherapy and radiation treatment with IMRT was initiated.
- Unfortunately, six months after undergoing definitive surgery, she experienced a relapse and passed away.

Surgical specimen after definitive surgery



Conclusion:

Relapsed patients have a far worse prognosis; as low as 10%. For the management of this uncommon tumour, new treatment approaches are therefore desperately needed.