

## Ewing's Sarcoma of Uterus – Case Report and Review of Literature

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### Introduction

Ewing's sarcoma is a rare bone cancer found in children and young adults. Extra osseous Ewing's sarcoma-Peripheral primitive neuroectodermal tumors (ES-PNET) are very rare. There are case reports of these in literature. We present a case of Ewing's sarcoma of the uterus.

### Case

A 22 years old woman with no significant medical or surgical history was admitted under the general surgeons for appendectomy. At laparoscopy, incidentally a small blood clot was found at the fundus of the uterus. Gynaecology opinion was obtained and it indeed looked like a 3 cm haematoma at the fundus from possibly a previous traumatic event. There was no instrumentation of the uterus done at surgery. As the patient was under general anaesthesia a recent history of any uterine surgery couldn't be confirmed. The patient made a good recovery from surgery and was seen in the gynaecology out patient with an ultra sound scan in 3 months. At that visit the scan showed that the haematoma had marginally increased in size. Patient denied any gynaecological surgery such as termination of pregnancy or hysteroscopy. A repeat scan was arranged in 2 months, as the patient was asymptomatic. The scan showed that there had been a rapid increase in size of the uterine lesion to 10 cms. An urgent MRI scan was performed and the case was discussed in the gynaecologic oncology multi-disciplinary meeting. The MRI showed a large mass at the anterior surface of the uterus measuring 10 cms and highly suggestive of sarcoma. The para aortic and pelvic lymph nodes were noted to be enlarged.

Patient had a laparotomy and a large friable mass from the anterior surface of the uterus was found. The disease was involving the omentum as well. Frozen sections taken were not conclusive but suggested possibly sarcoma or lymphoma. The patient had total abdominal hysterectomy, left salphingo oophorectomy, right salpingectomy, omentectomy and pelvic and para aortic lymphadenectomy. Right ovary was conserved. She made a full recovery from the surgery.

The histology showed, round blue cells as well as small spindle cells with increased vascularity and a very high mitotic count (Figure 1).

The tumour was positive for CD99, CD117 and Beta-catenin. A provisional diagnosis of High-grade endometrial stromal sarcoma was made and the specimen sent for second opinion. On review 2 differentials were suggested: The first was a tumour in the Ewing family (peripheral primitive neuroectodermal tumour). The second was the specific variant of high-grade endometrial stromal sarcoma associated with YWHAE-FAM22 gene re-arrangement. It was suggested for a molecular analysis to look for the translocations associated with both Ewing family of tumours and the high-grade endometrial stromal sarcoma. Molecular testing showed FISH positivity for EWSR1 rearrangement along with a PCR transcript showing type 1 ESWR1-FLI1 translocation indicative of Ewing's sarcoma. The omentum was positive for the tumour. The nodes were negative.

Patient will be soon starting chemotherapy and possibly radiotherapy.

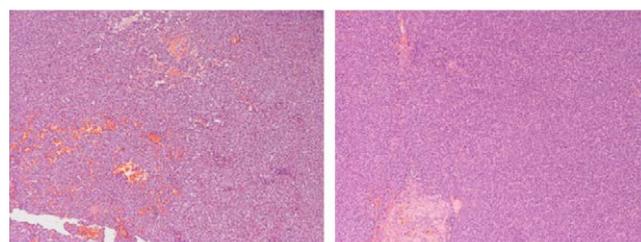
### Discussion

Primary Ewing's sarcoma-primitive neuroectodermal tumor (ES-PNET) of the uterus is an extremely rare malignancy.

ES-PNET is a group of tumors thought to be derived from fetal neuroectodermal cells that has morphologic features of small round cell tumors with variable degrees of neural, glial, and ependymal differentiation [1]. ES-PNET is mainly of two main categories - central and peripheral. Central ES-PNETs are derived from the neural tube and mainly involve the brain and spinal cord. Peripheral ES-PNETs are derived from the neural crest and occur outside the central nervous system, often involving the sympathetic nervous system or soft tissue and bone [2].

There have been case reports of ES-PNET occurring in extraosseous tissues like breasts, liver, kidneys, Ureter etc. In female genital tract, there are case reports of their origin from vulva, vagina, cervix, ovary and Uterus [3,4].

ES-PNET of uterus if found is usually in young adults or postmenopausal women. Abnormal vaginal bleeding is the commonest symptom, however they can present as pelvic mass or they can be asymptomatic and found incidentally as in our case. These are very aggressive tumours and are usually advanced by the time of diagnosis. At diagnosis there is usually macroscopic metastasis in 25% cases while most have micrometastasis [3]. Pre-operative diagnosis is very rarely considered due to the rarity of these tumours. The diagnosis is usually



**Figure 1:** The histology showed, round blue cells as well as small spindle cells with increased vascularity and a very high mitotic count.

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made on histology. Routine histology and immunohistochemical examinations are sufficient to make a diagnosis. However at times genetic analysis is needed [4]. The differential diagnoses are lymphomas, clear cell sarcomas and other sarcomas with neural elements [1].

All reported cases except one were managed mainly by surgery in the form of hysterectomies followed by chemoradiotherapy. One case was managed by chemoradiotherapy alone [1]. Burchil suggested advance age, tumour size more than 8 cms and Pre-treatment elevated LDH as poor prognostic features [5]. The overall prognosis was poor in these reports due to late presentation and aggressive nature of the tumour. However a review published in 1997 compared 130 paediatric patients with extraosseous ES-PNET and 2792 paediatric patients with rhabdomyosarcoma. All these patients were treated with surgery, chemotherapy and radiotherapy. Response to treatment was found to be similar with 80% response rate and 5 years survival 70-75% [6].

ES-PNET are very rare and aggressive tumours which are found mainly in adolescent and postmenopausal women. Multi modal management in the form of surgery and chemoradiotherapy is needed. In spite of this, the overall prognosis is variable.

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