

Ewing's sarcoma of the uterus in a 23-year-old female

A case report

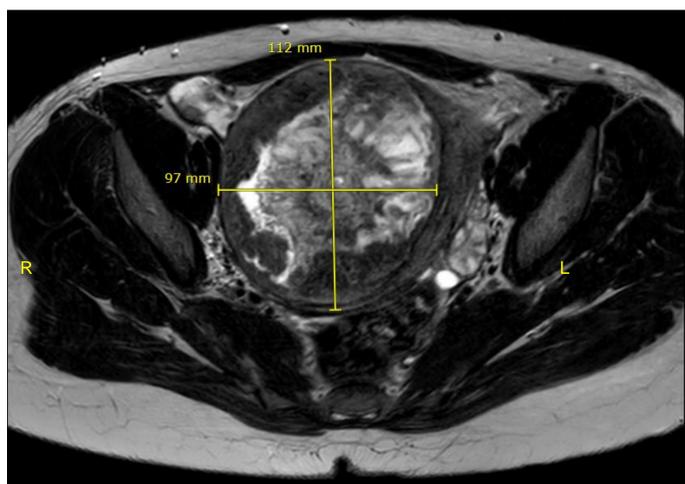
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Introduction

Extraskelletal Ewing's sarcoma of the uterus is very rare as less than 50 cases were published in English literature at the time of diagnosis.

Case report

A 23-year-old nulliparous female presented herself with an increased abdominal volume and lower abdominal pain. Gynecological examination revealed a large tumor in the uterine wall, interpreted as a myoma. The patient was treated with Esmya® for 3 months, in an attempt to reduce the myoma volume. The follow-up examination unexpectedly showed further growth of the tumor. An MRI revealed a 10 x 11 cm inhomogeneous and contrast enhancing mass related to the anterior and side wall of the uterus.

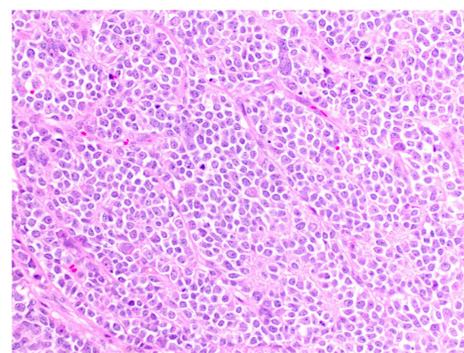


MRI of the pelvis: intramural, inhomogeneous, contrast enhancing tumor of the uterus (10 x 11 cm)

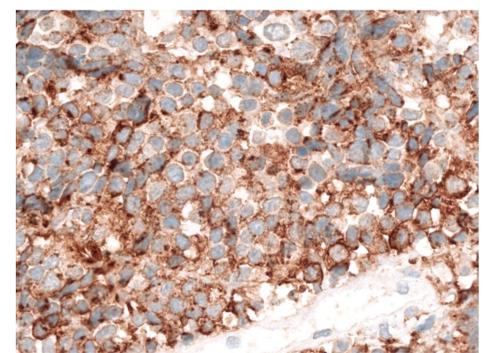
In a private hospital, a biopsy of the tumor was performed. Histological examination revealed an extraskelletal Ewing's sarcoma/primitive neuroectodermal tumor (PNET) with a high proliferative MIB-1 index of 80%. The following PET-CT showed persistent, partly necrotic tumor of 11 cm in diameter, but no distant metastases.

There is evidence for the impact of multidisciplinary and multimodal treatment of patients with Ewing's sarcoma.

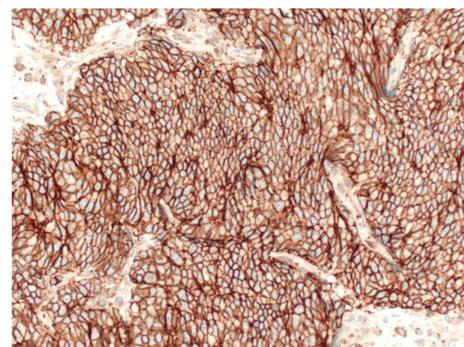
The treatment approach was based on experience for Ewing's sarcoma of the bone. Our patient underwent primary chemotherapy with six cycles of VIDE. The tumor showed regression with a residual tumor mass of 4 cm. We performed a median laparotomy with hysterectomy, bilateral salpingectomy and lateral transposition of the ovaries. On histology of the uterus the tumor showed 80% necrosis, however 20% vital tumor areas remained.



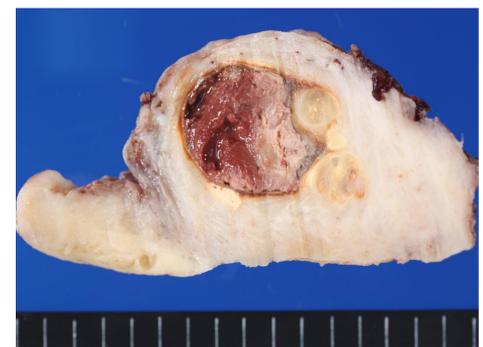
Ewingsarcoma: sheets and nests of a small, blue and round cell tumor (HE, 200x)



The tumor cells express CD57 (CD57, 400x)



Typical membranous CD99 expression of tumor cells (CD99, 200x)



Uterus: Lobulated tumor mass with extensive necrosis (red-brown) and smaller nodules of viable tumor (yellowish, right side of tumor)

After surgery, the patient received adjuvant treatment with radiotherapy of the pelvis (cumulative dose of 45 Gray) and chemotherapy with VAC. On follow-up the CT showed a complete remission. The patient presented in a good general state of health. During the entire treatment, the GNRH agonist Zoladex® was applied to achieve ovarian suppression and to prevent chemotherapy-induced gonadotoxicity and premature ovarian failure.

Discussion

In spite of its rarity, Ewing's sarcoma should be included in the differential diagnosis of a growing uterine tumor, especially in adolescent and postmenopausal women, as most of the cases have been reported in those 2 age groups. There is no consensus on the adequate treatment, due to the lack of studies. Multidisciplinary management of this rare disease is key and multimodal therapy consisting of surgery, chemo- and radiotherapy improves the prognosis, which is still poor with an overall 5-year survival rate of 50%.