MASSIVE AND EXTENSIVE PNET OF THE UTERUS DURING PREGNANCY: MANAGEMENT

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CASE REPORT: INTRODUCTION

In this case report we describe the obstetric and oncological outcome of a huge mass diagnosed as a leiomyoma in a woman of 39 years pregnant at 22 weeks of gestation who complained of low back pain, dysuria and urinary frequency. During the 25th week, the patient came to our attention at night, with severe anemia and suspected haemoperitoneum.

MANAGEMENT

1° SURGERY AND PARTUM

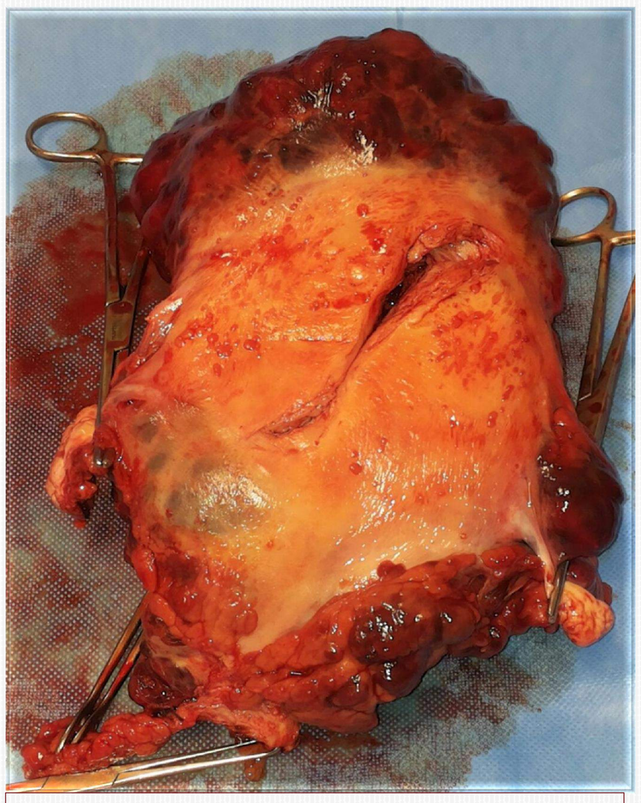

Figure 1 Uterus with sagittal cesarean cut on the posterior face and adnexa.

She underwent an emergency cesarean section, with the extraction of the fetus, alive and well, weighting 400 g.
During surgery, we surprisingly found a uterine sarcoma-like metastic tumor.

We made hysterectomy **(fig. 1),** bilateral salpingo-oophorectomy, pelvic peritonectomy, omentectomy, appendectomy and excision of some bulky lymph nodes.

STAGING

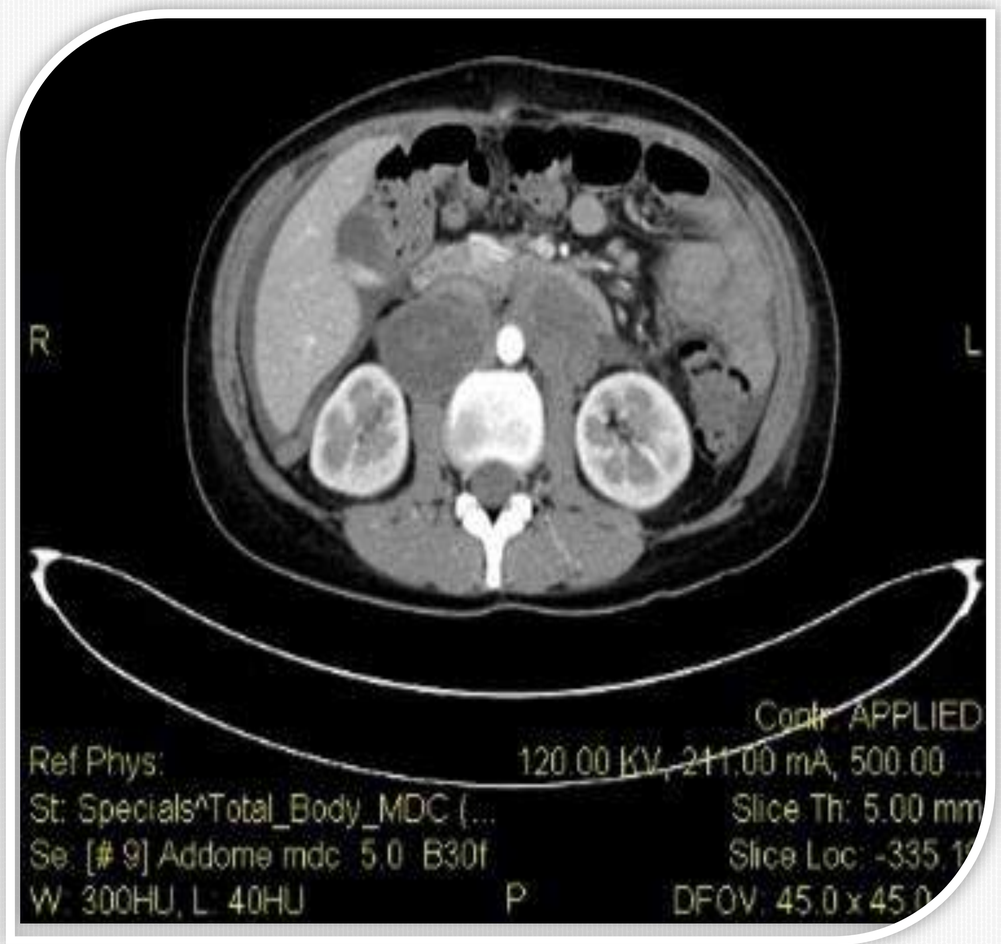

Figure 2

CT scan of the abdomen, after emergency surgery, revealed bulky lymph nodes that compressed inferior vena cava causing its focal thrombosis from kidney level till the right common iliac vein.

DIAGNOSIS

- Histological examination revealed a uterine body PNET (peripheral primitive neuroectodermal tumor). PNETs are a family of highly malignant neoplasms characterized by small round cells **(fig.3)** of neuroepithelial origin.
- They usually involve bone and soft tissues with a higher incidence in childhood.
- This is the first case with onset in uterine body during pregnancy.

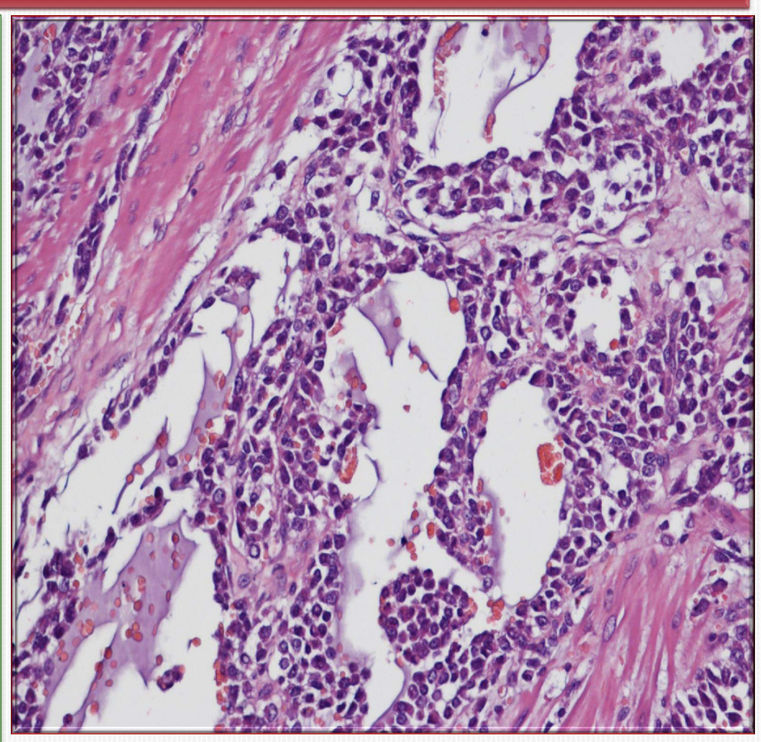

Figure 3 Small round cells udifferentiated tumor invading miometrium creating nets with diffuse lymphovascular space invasion.

This image was kindly granted by Dr. Maria Grazia Fiore, Department of Pathology, University of Bari, Italy.

2° SURGERY

So we completed debulking with a second surgery **(fig. 4)** including diaphragmatic peritonectomy and excision of a huge lymph node from lombo-aortic lymphadenectomy.

We also dissected a mass infiltrating the left renal vein through its interruption and reconstruction.

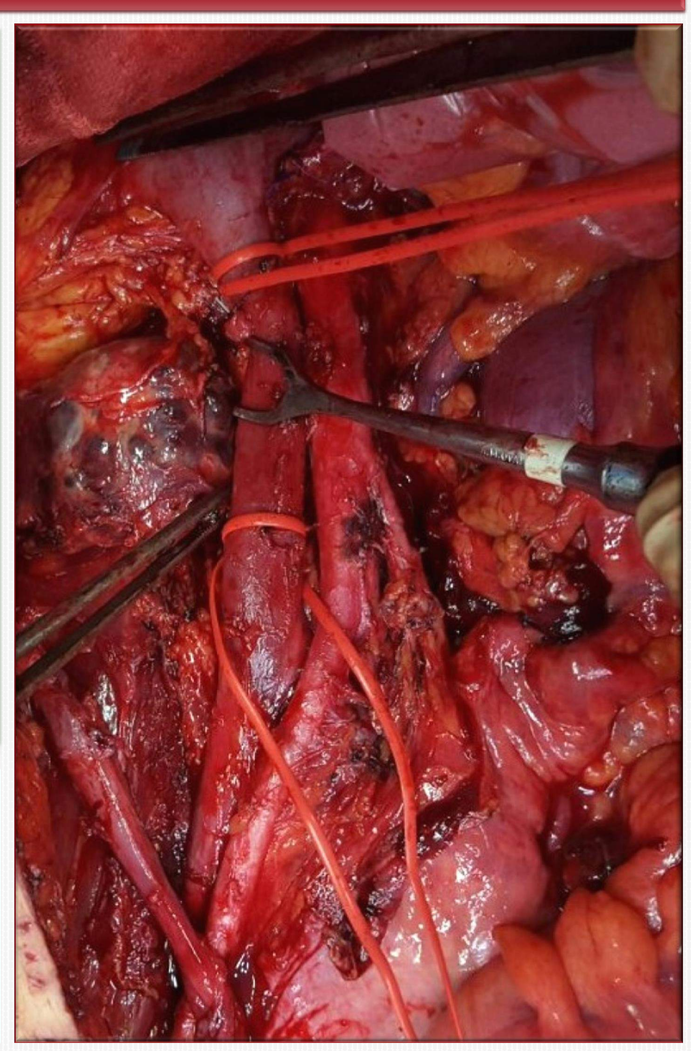

Figure 4
From the right side we find retrocaval bulky lymph node under renal veins level, isolated inferior vena cava and aorta.

CONCLUSION

After a group discussion, we decided to refer the patient to chemotherapy with doxorubicin, etoposide and ifosfamide added to the classic cyclophosphamide-vincristine-actinomycin regimen.

CHEMOTHERAPY

Ten day after second surgery, echo-colour Doppler showed a regular microcirculation in left kidney.
The patient was discharged after 10 days, while the baby after a month, both in good health.