

METASTATIC PRIMARY OVARIAN PEComa: A CASE REPORT

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PERIVASCULAR EPITHELIOID CELL TUMOR (PEComa)

- Rare mesenchymal neoplasm, characterized by expression of both melanocytic and smooth muscle markers. ^[1]
- Potential precursor lesion: minute perivascular epithelioid cell nests occurring in lymph nodes. ^[2]
- Associated with tuberous sclerosis complex (TSC) and frequently harbour TSC1/2 mutations including genetic pathway Rheb/mTOR/p70S6K (responsive to mTOR inhibitors). ^[3,4]
- Usually benign but some cases are malignant. ^[5]
- Common origin: abdominopelvic viscera especially GI and uterus. ^[6]
- Gynaecologic PEComas: uterine corpus, cervix, vagina, broad ligament, vulva and adnexa. ^[7]
- Ovarian PEComas: metastases of PEComa to the ovary, PEComatosis involving the ovary, ^[7] and primary ovarian PEComas (3 cases in literature). ^[8-10]

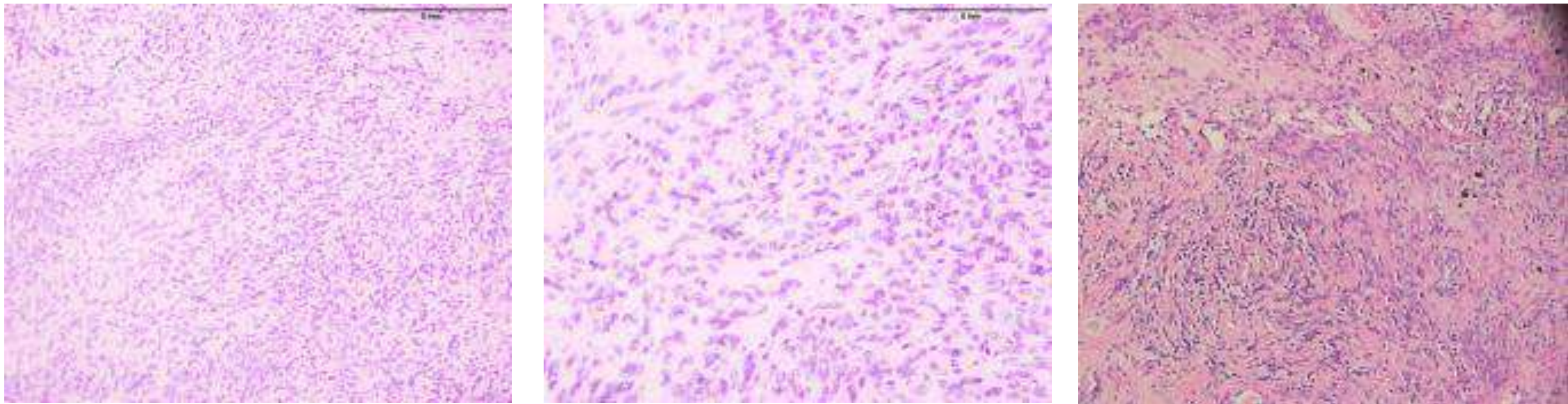


Figure 2: Microscopic Pictures of Primary Ovarian PEComa: (a) HMB-45 positive and (b) SMA positive

CASE SUMMARY

- A 31-year-old female, married for 4 years, para 1 with 2 miscarriages and no comorbidities underwent excision of left adnexal mass outside SKMCH. Biopsy showed Perivascular epithelioid cell tumour (PEComa).
- Follow-up serial scans (figure 1): Residual complex solid cystic mass in the left adnexa with interval regression in cystic and solid components.
 - After MDT discussion at SKMCH, she underwent fertility-sparing comprehensive surgical staging i.e., left salpingo-oophorectomy, excision of left pelvic wall mass excision, and infracolic omentectomy.
 - Biopsy (Figure 2) showed left ovarian primary PEComa with metastatic deposits in the omentum and left pelvic sidewall.
 - Case re-discussed in MDT: Targeted therapy is being planned after completion of pregnancy.

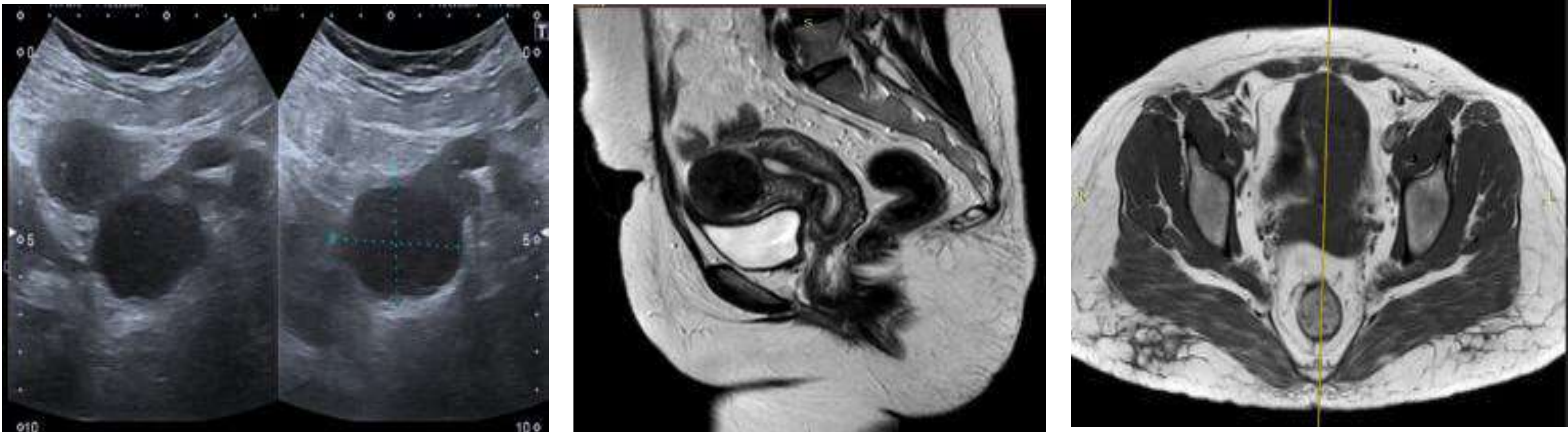


Figure 1: (a) Ultrasound Images (b) MRI Pelvis with IV contrast (Axial and Sagittal Sections)

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