Introduction
Epitheloid sarcoma is rare and aggressive malignant soft tissue tumor. 1 Epitheloid sarcoma of vulva is even rarer and only 20 cases have been reported. 2

Case Report
A 46 year old female, presented with a palpable, firm, non tender mass of right labia majora. USG of the abdomen and pelvis revealed multiple submucosal (2.9cm), subserosal (2.1cm) and cervical fibroids. Right ovary was normal and left ovary had multiple simple cysts, largest measuring 2.3cm. She underwent Total abdominal hysterectomy bilateral salpingo-oophorectomy with wide local excision of the labial mass.

Histopathological report suggested high grazed malignant epithelial tumor of the labia. Immunohistochemistry showed AE1 positive, EMA positive, Vimentin Positive, AFP negative, Desmin negative, CK-7 negative and was concluded to be Epitheloid sarcoma, Proximal type. A thorough metastatic work up was done and was found to be normal. The patient was delivered postoperative radiotherapy 50Gy in 25 fractions five day a week over five weeks.

Result
Patient is currently on follow up and is asymptomatic.

Keywords
Epitheloid sarcoma, vulva, proximal type.
The primary treatment of epitheloid sarcoma of vulva consists of wide local excision of the lesion with an aim of achieving 2 cm margin. The role of adjuvant radiotherapy is controversial. One study reports a recurrence rate of 14% in patients who received adjuvant radiotherapy as compared to 71% in patients who did not receive it (p=0.01). Another study recommends adjuvant radiotherapy for high grade tumors and those with inadequate margins. The role of chemotherapy in this condition is an adequate. A variety of chemotherapy regimens have been employed but without any significant results and chemotherapy for recurrences is even less effective. Overall five year survival rate for distal type of epitheloid sarcoma is 50 to 80% and 10 year survival rate is 42-45% but proximal type has poor prognosis.

Indicators of poor prognosis of epitheloid sarcoma include vascular invasion, depth of invasion more than 0.5 cm, tumor size more than 5 cm. more than 30% necrosis, high mitotic activity, nodal involvement, multifocal local disease, presence of local recurrence and advanced stage at presentation, male sex, older age at presentation. Two variety of epitheloid sarcoma are described in literature. “Proximal type” type variant first described in 1997 and occurs in perineum and pubic region and have a distinct entity than “distal type”. Distal type occurs in upper and lower extremities. Proximal type epitheloid sarcoma have higher rate of local recurrence and distant metastasis. Local recurrence is common and may develop after many years post successful treatment, despite adequate and negative resection margin. Distant metastasis eventually occurs in up to 60% of cases. Excision with wide surgical margins is the treatment of choice. Surgery is also recommended for local recurrences. Removal of inguinal lymph node is controversial. Dissection of loco-regional lymph node is considered if there is suspicious or enlarged node at physical examination or in imaging studies. Unlike most soft tissue sarcoma, epitheloid sarcoma characteristically spread to lymphatics. 34% patient presents with lymph node metastasis at the time of diagnosis. Post operative radiotherapy decreases the recurrence rate in case of close margin or high grade histology the role of adjuvant chemotherapy is minimal even in metastatic disease. 5 year and 10 year survival of epitheloid sarcoma is 50-70% and 42-55% respectively. Optimal treatment of epitheloid sarcoma of vulva is not known due to its rarity.

**REFERENCES:**