

# Case of the month

## April 2021

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Senior Consultant

- A 45-year-old diabetic and hypertensive female presented as a case of ?bilateral adnexal masses
- She was investigated elsewhere, where PET scan revealed a heterogeneously enhancing large pelvic mass

# PET scan



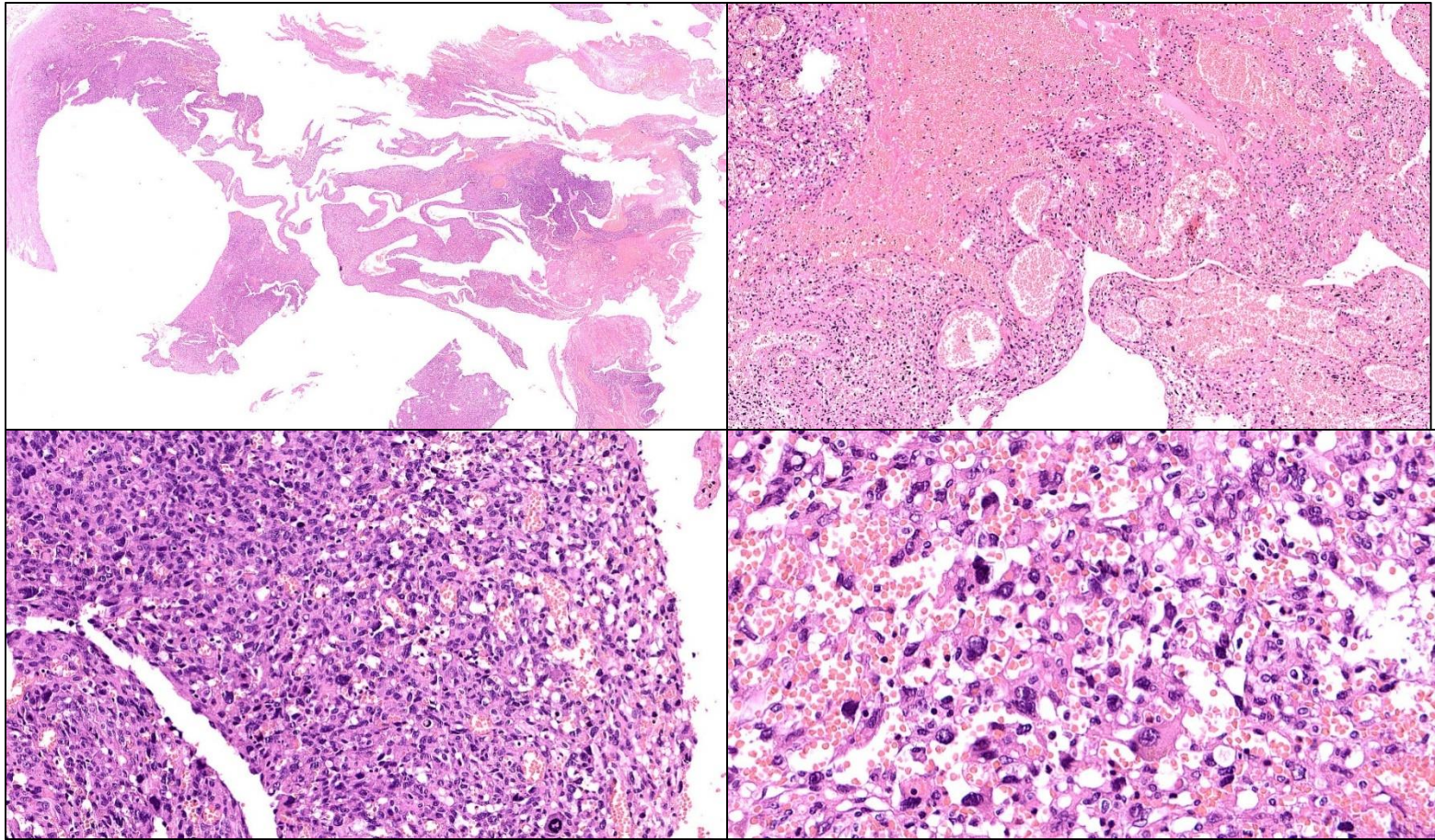
- She underwent Total abdominal hysterectomy with bilateral salpingoopherectomy and omentectomy outside our institute
- Sections of the ovarian tumor were submitted to our deptt for a second opinion and IHC
- The gross images were provided by the primary reporting pathologist
- Acknowledgement Dr Tanu Aggarwal

# Gross

- Right ovarian mass measured 16cm in size
- Cut section:
  - Solid cystic
  - Dark brown
  - Showed multiple variable sized cysts filled with hemorrhagic fluid

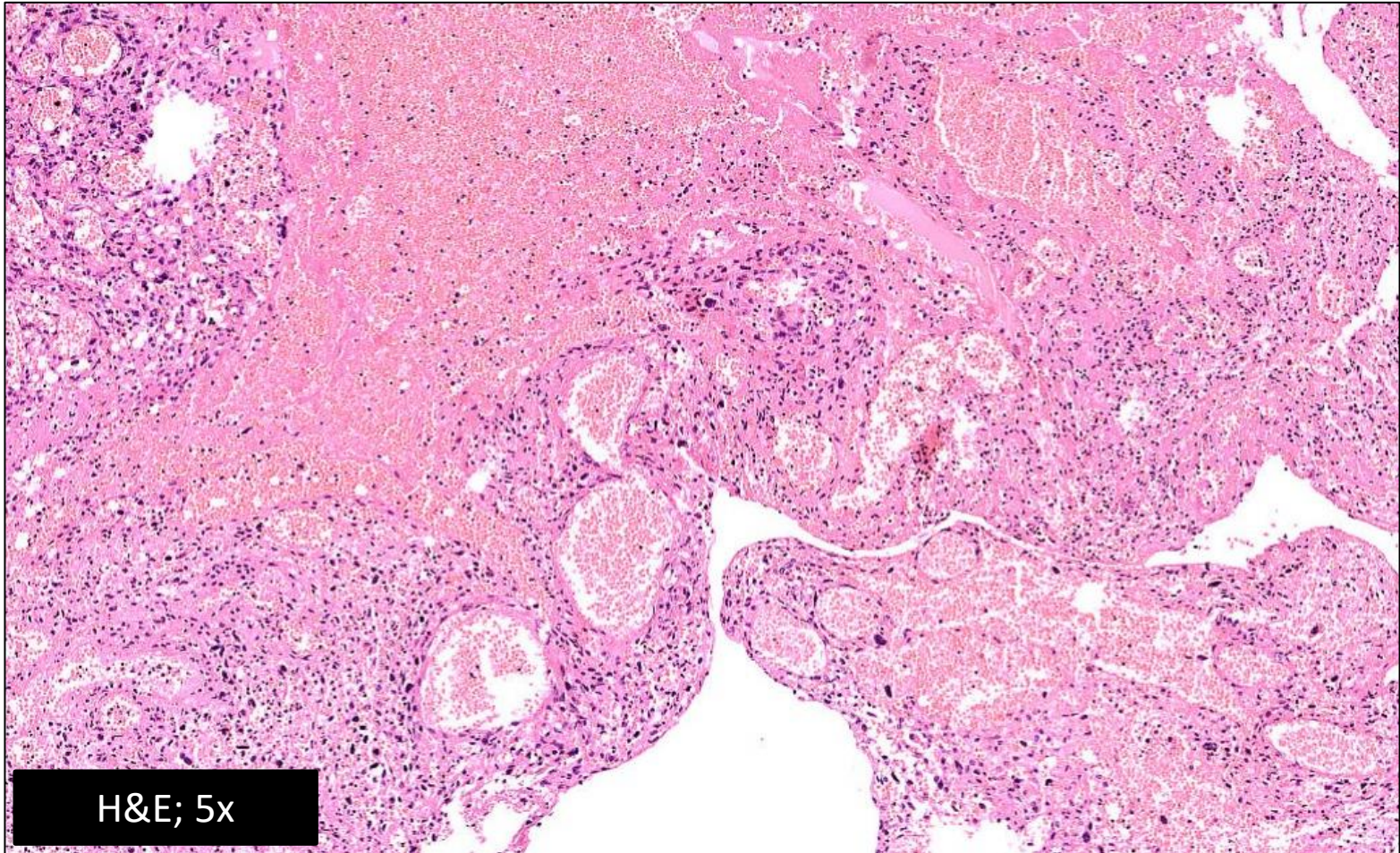


# Birds eye view of Microscopy

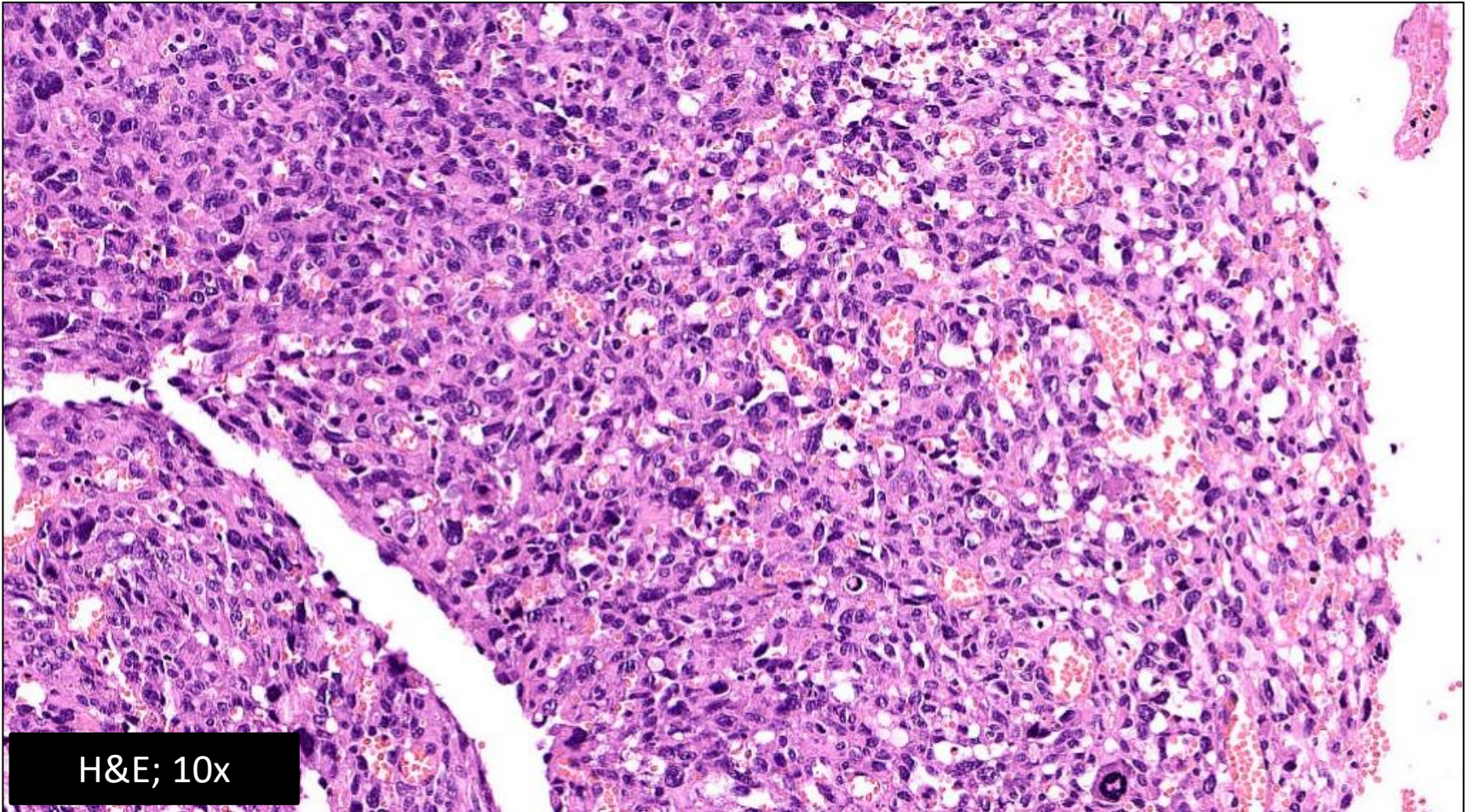


Diagnosis ??????

# Microscopy

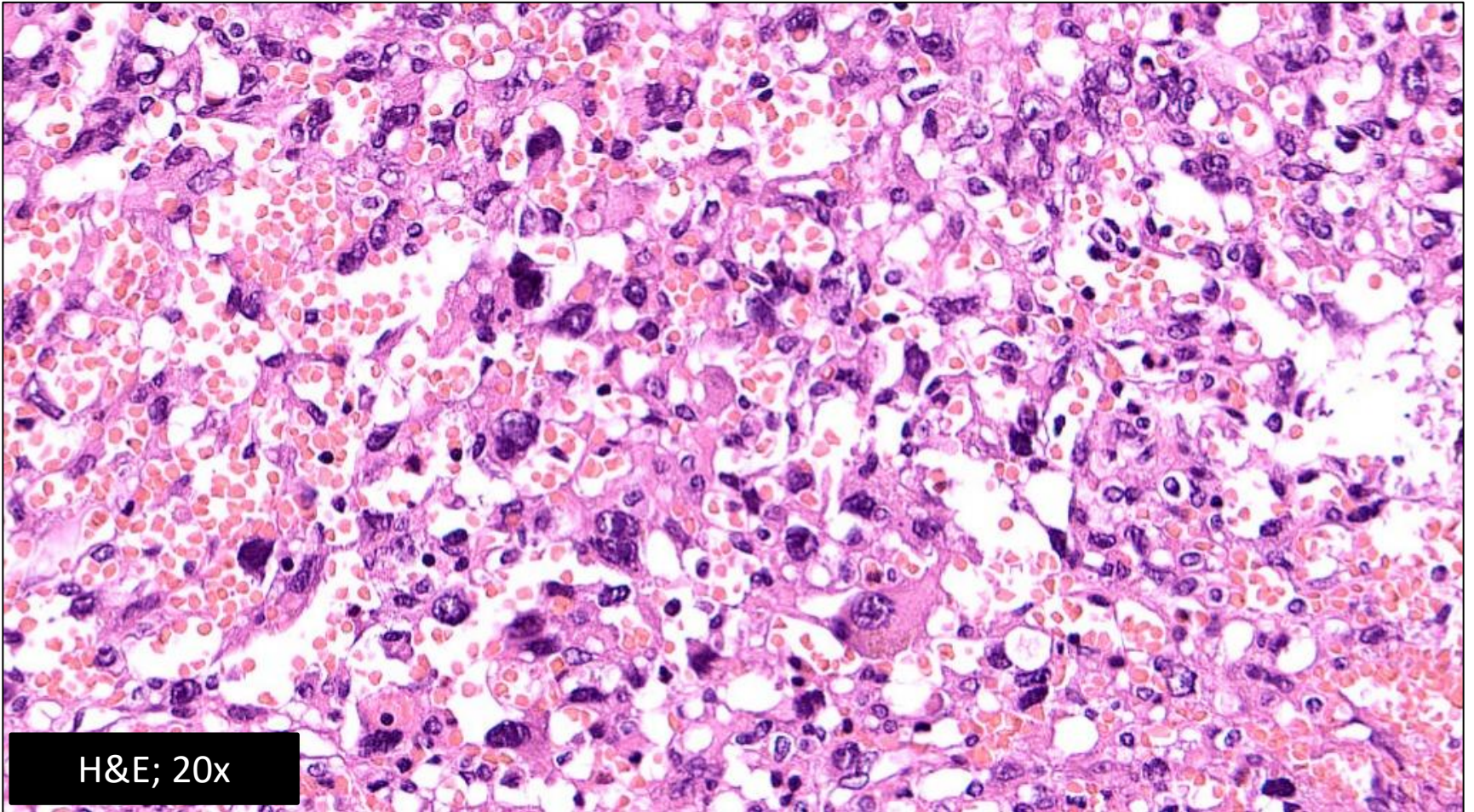


Sheets of tumor cells with blood filled spaces

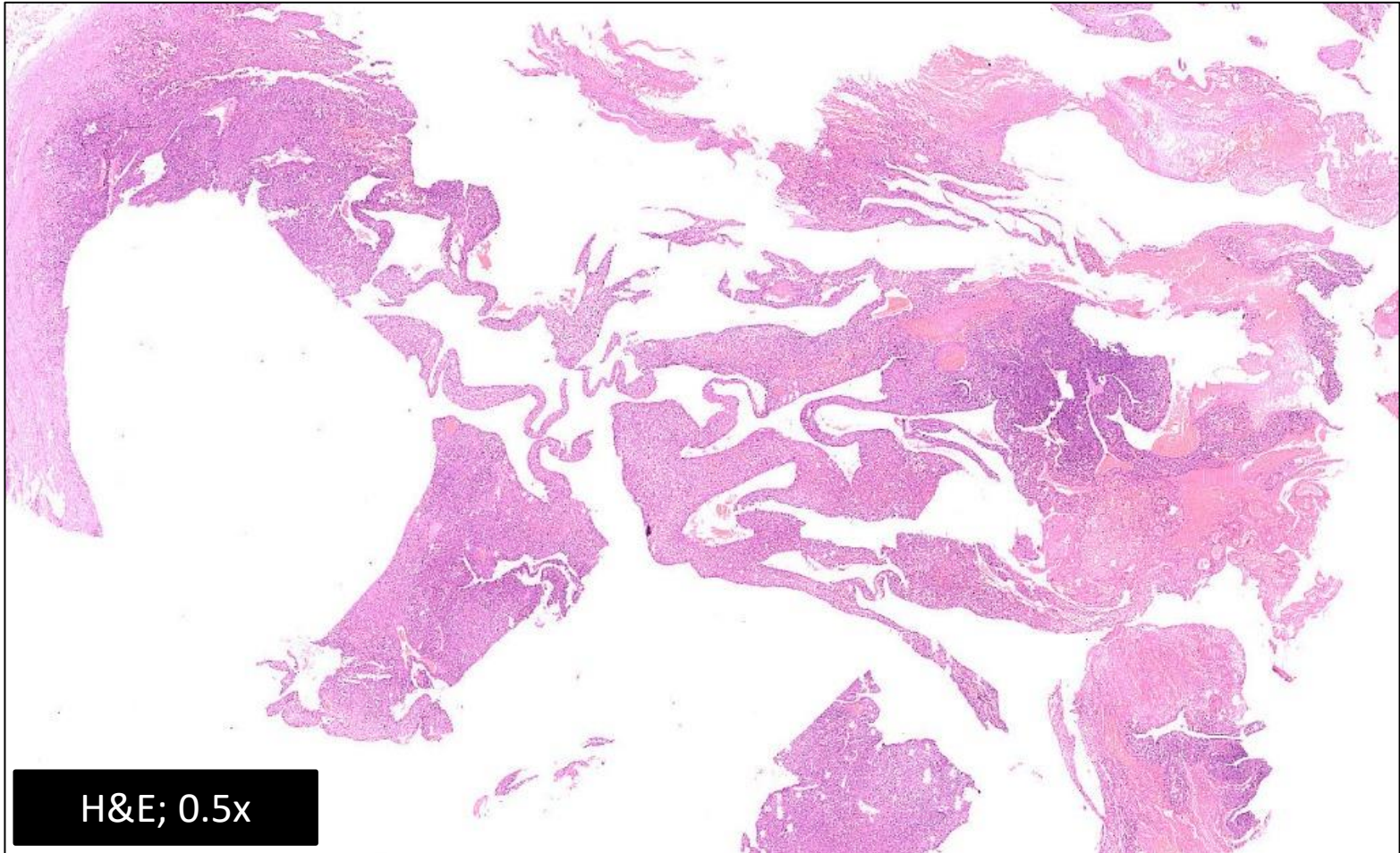


Cellular tumor showing neolumina formation containing RBCs

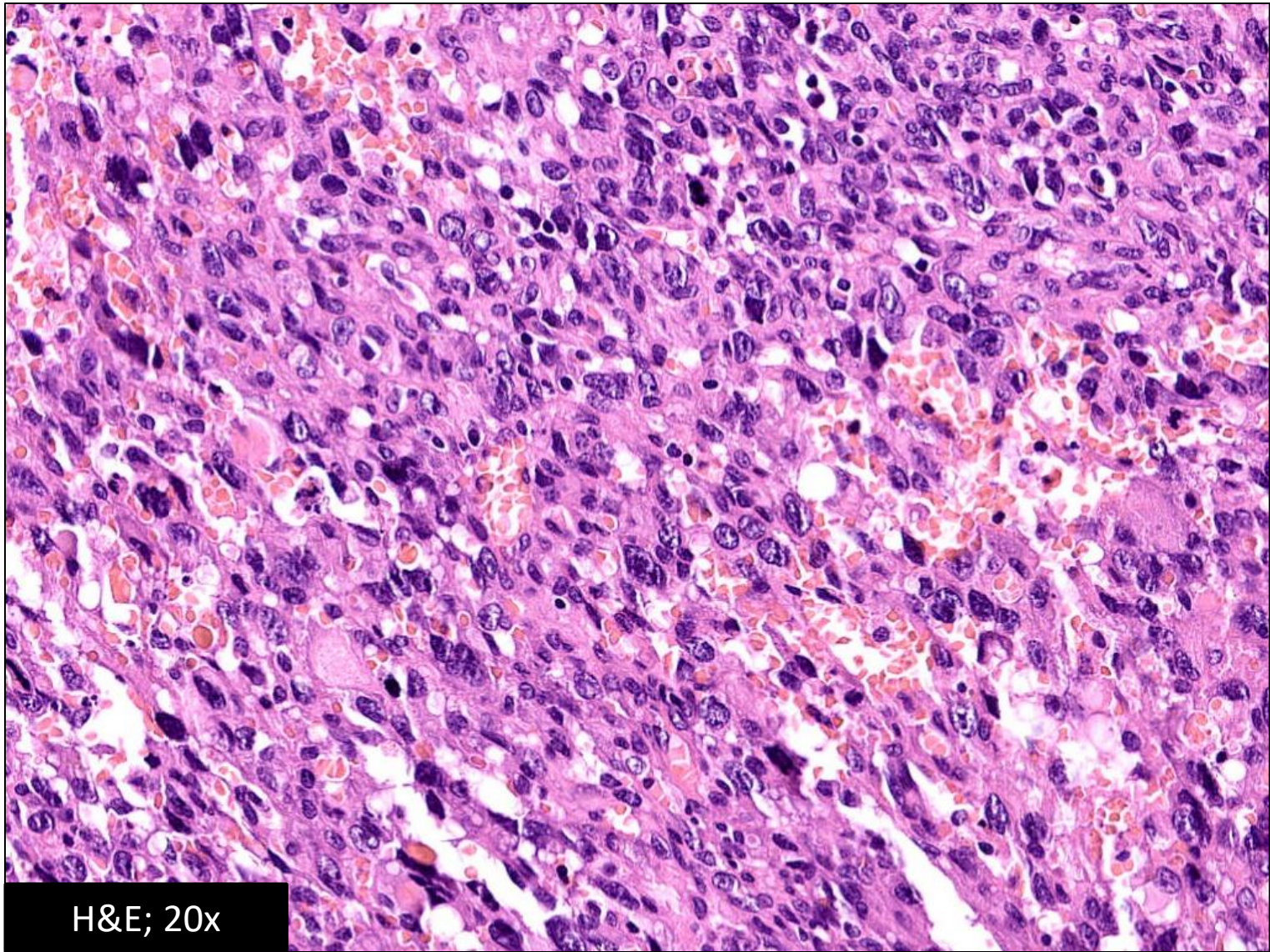




Tumor cells have markedly pleomorphic nuclei and eosinophilic to clear cytoplasm



Low power view shows anastomosing vascular channels



H&E; 20x

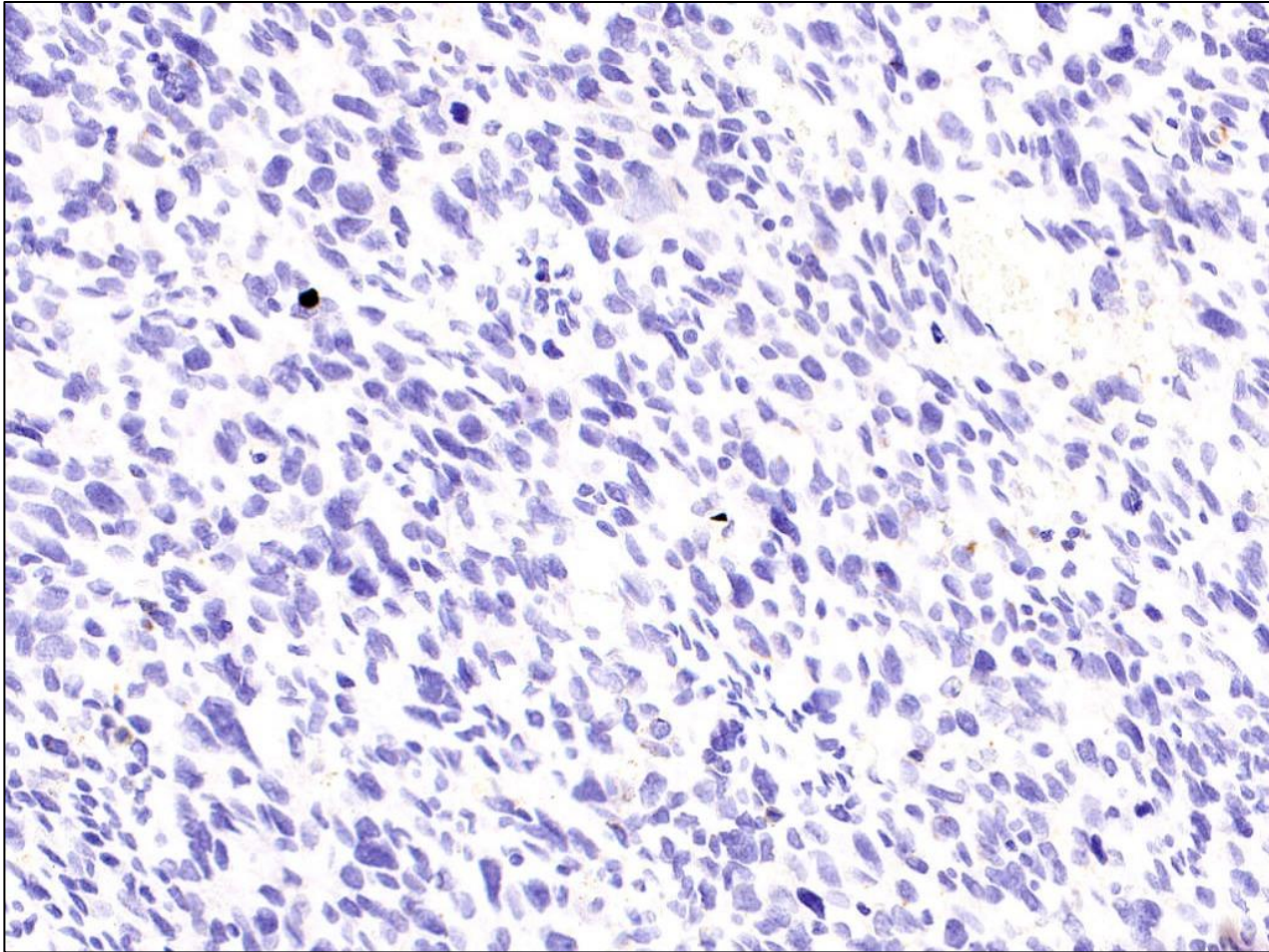
Nuclear anaplasia is stark, with brisk mitosis

# Differentials

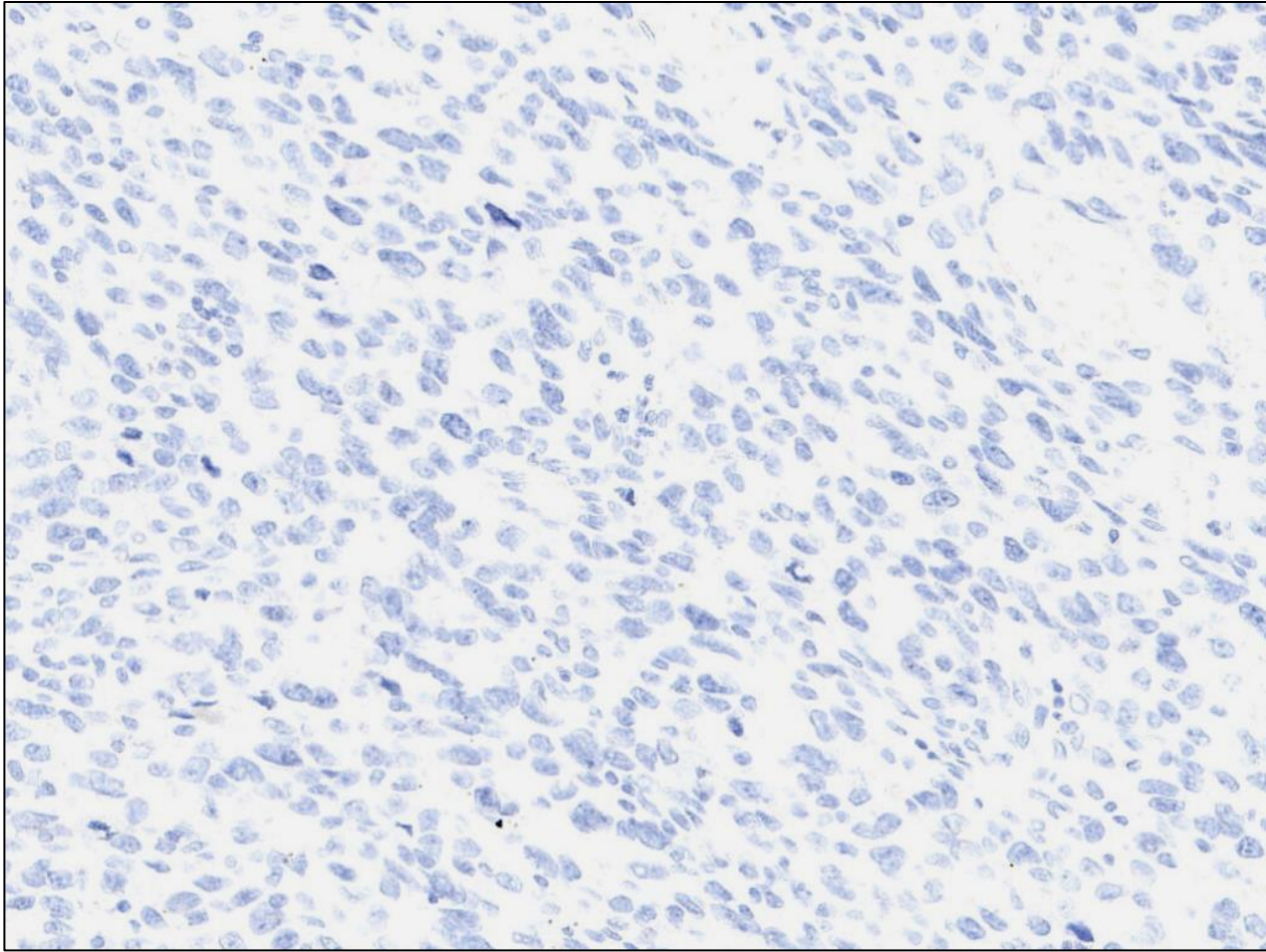
- Carcinosarcoma
- Undifferentiated Carcinoma
- Somatic malignancy in Teratoma
- Malignant mesenchymal tumor  
(smooth/skeletal muscle)
- Malignant vascular tumor

- Extensive sampling from tumor was done, which did not reveal any other component of the tumor nor did it show any synchronous tumor within the ovary
- Tumor was subjected to Immunophenotyping

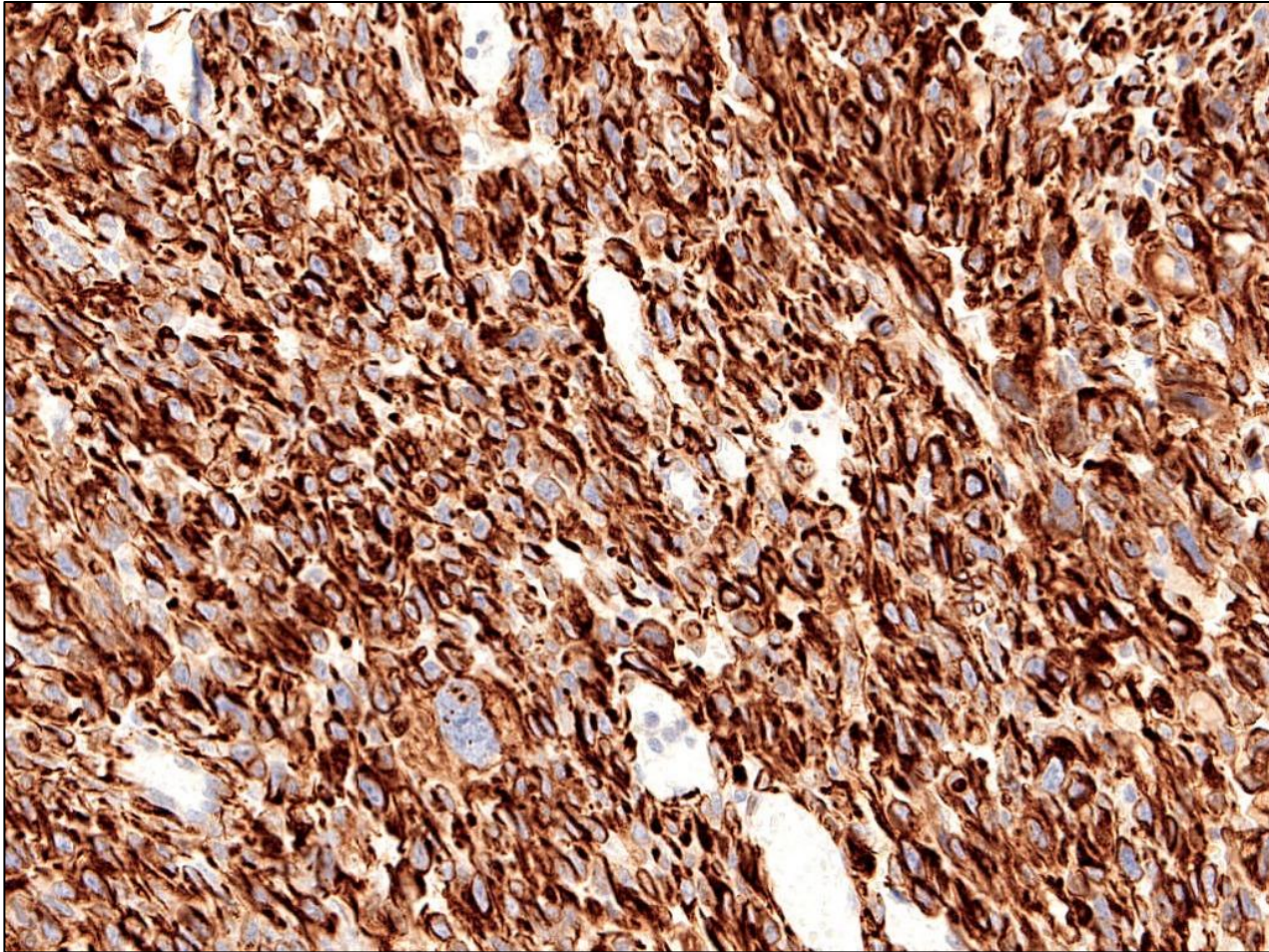
# CK: Negative



# PAX8: Negative

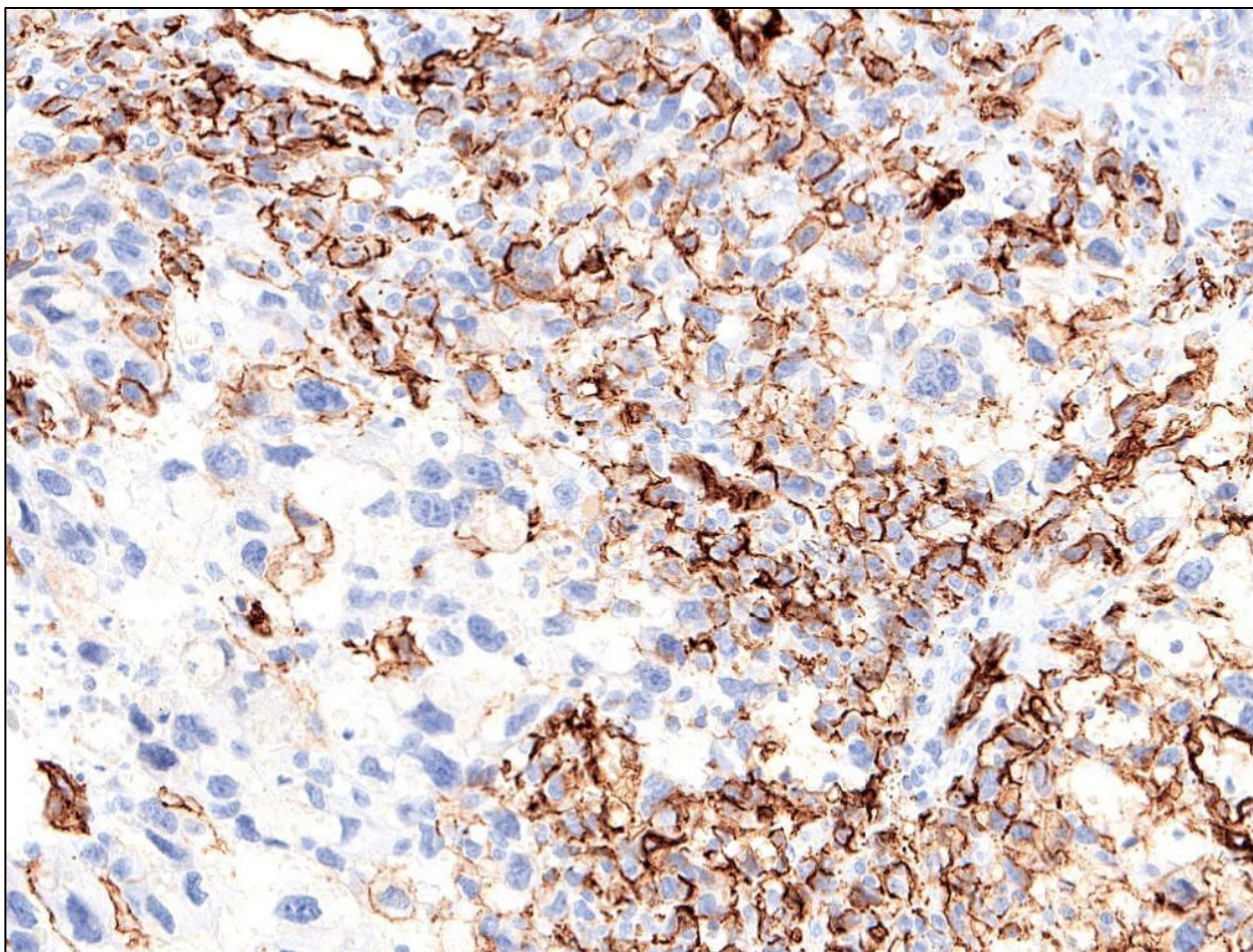


# WT1: Negative

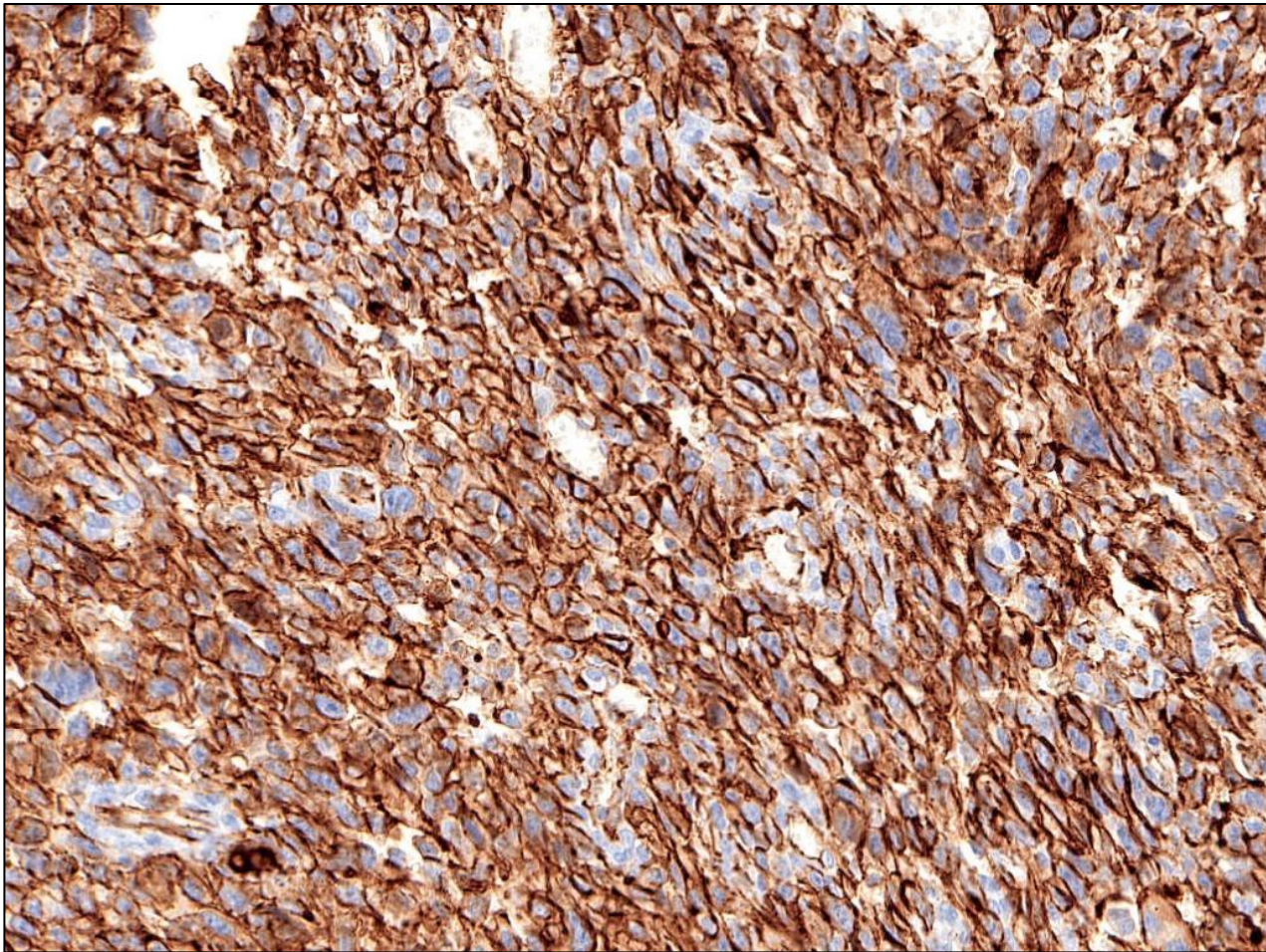




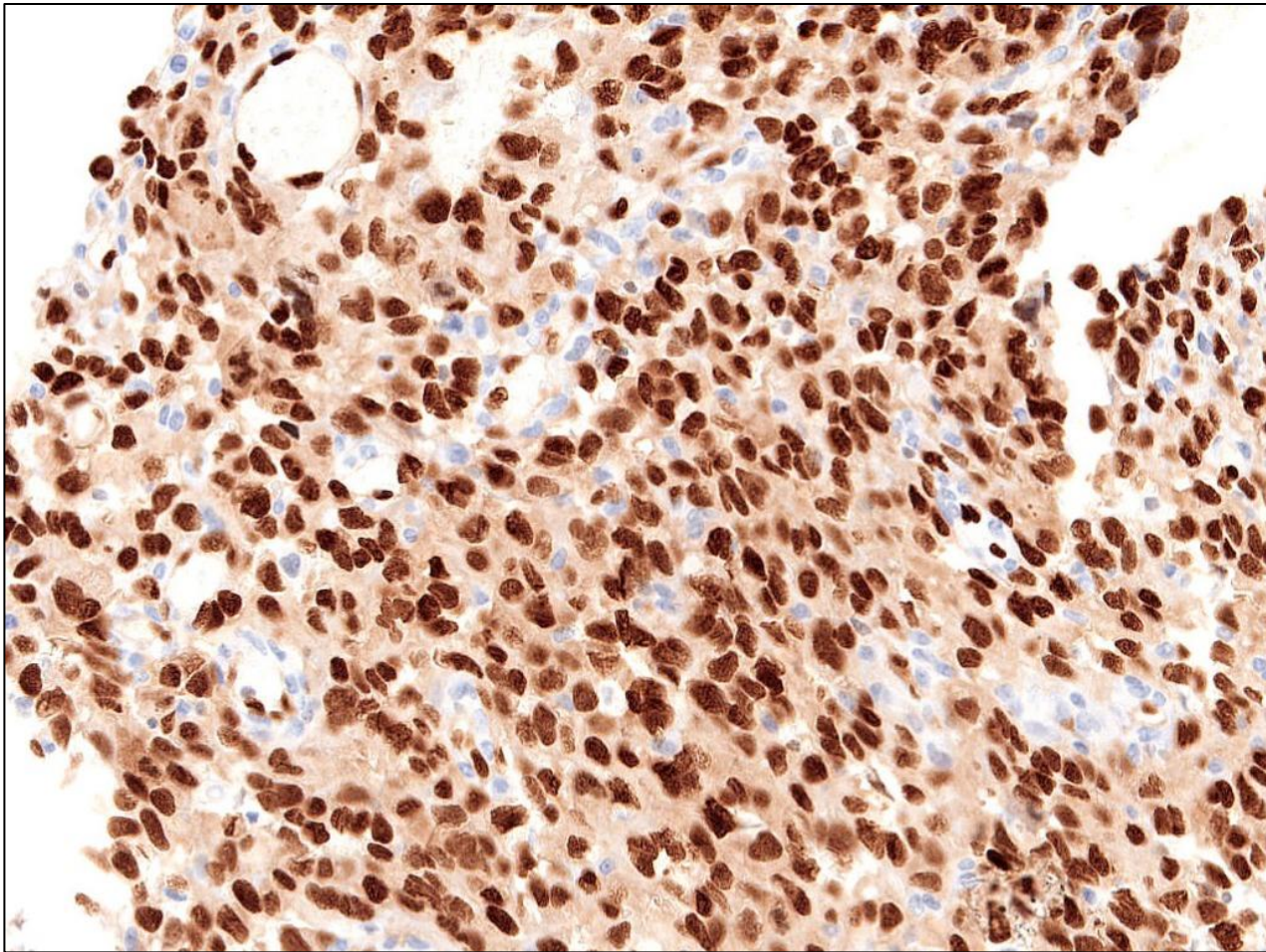
# CD34: Focal



# CD31: Positive



# ERG1: Positive



# Final Diagnosis

- High grade pleomorphic sarcoma of vascular lineage favouring

“Primary Angiosarcoma of ovary”

# Treatment plan

- Case was discussed in MSC and patient was advised for Paclitaxel & Carboplatin based chemotherapy
- Patient was started on chemotherapy (Currently post 3 cycles)

# Discussion

- Sarcomas of the ovary are rare tumors, comprising less than 1% of ovarian malignancies
- Angiosarcoma can occur as a pure sarcoma or in combination with ovarian tumors such as Teratoma, mucinous cystadenocarcinoma and dermoid cysts

- The incidence of primary ovarian angiosarcoma is 1/1,000,000 of ovarian malignant tumors
- Only 32 cases of primary ovarian angiosarcomas have been described in the previous literatures
- They are more common in the young and premenopausal women

- Histopathological confirmation is essential for the final diagnosis of primary ovarian angiosarcoma
- It is often characterized by vasoformative arborizing channels of variable sizes and shapes lined by atypical endothelial cells
- Mitosis is generally high
- Necrosis may be seen



# IHC

- Tumors typically stain for vascular markers such as
  - CD31
  - CD34
  - ERG1
  - factor VIII
- while are characteristically negative for CK

# Treatment options

- Surgical debulking
- Post-operative adjuvant chemotherapy
- Radiotherapy

# Conclusion

- Ovarian angiosarcoma is a very rare tumor with poor prognosis for patients with advanced stage.
- The diagnosis of angiosarcoma is highly relied on the identification of communicating and typical vascular-like structures, with positivity of specific endothelial IHC markers being a diagnostic prerequisite.
- Complete surgical resection and postoperative adjuvant chemoradiotherapy are routine treatment methods.

THANK YOU