

# Case of the month

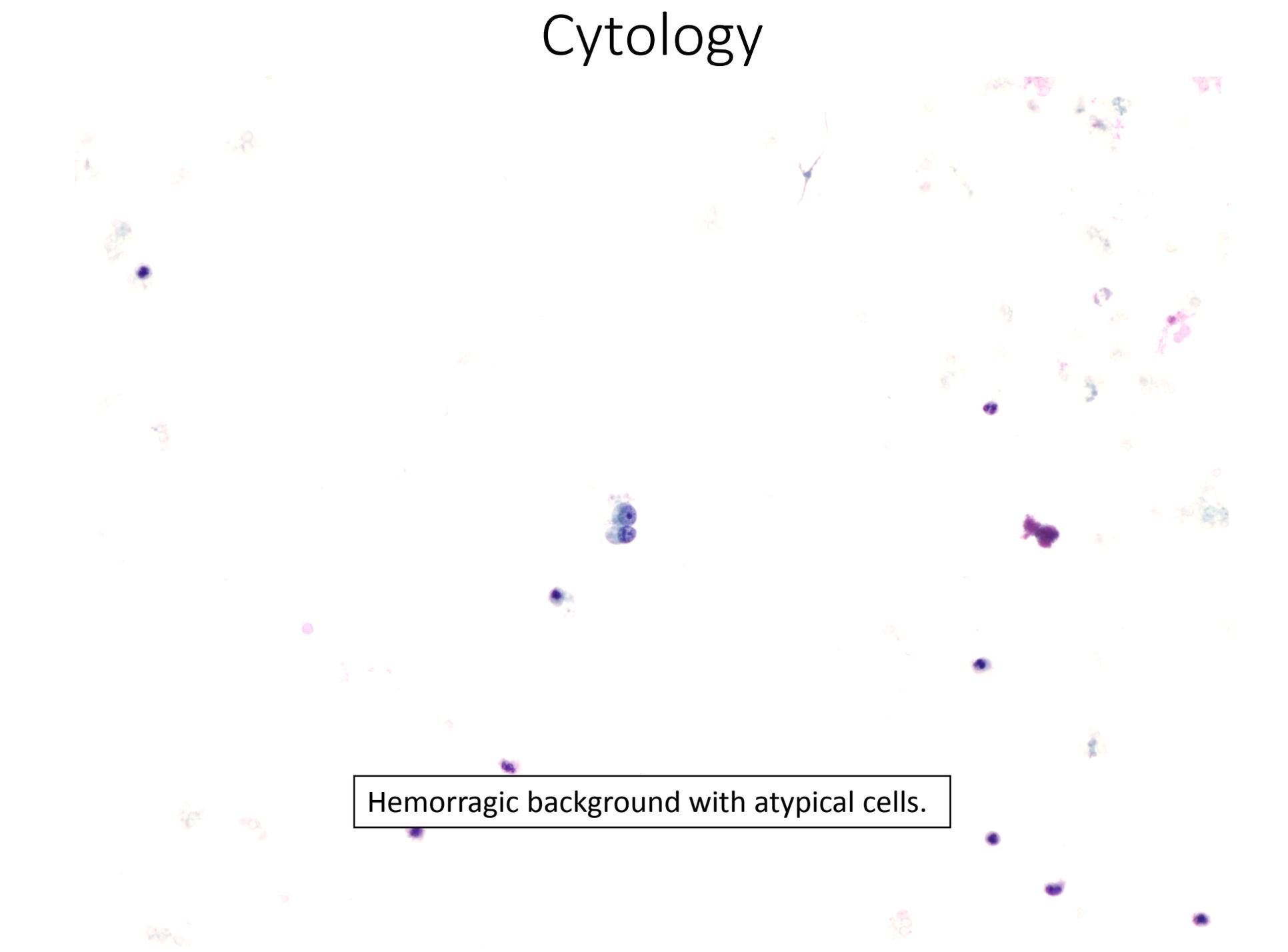
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# Clinical history

- A 56-year-old menopausal woman
- Admitted to gynecologic emergency for vaginal bleeding
- No history of cervical carcinoma
- A Pap smear was performed at emergency

# Cytology

A cytology slide showing a hemorrhagic background with atypical cells. The slide is stained with a combination of pink and purple dyes. The background is a light pinkish-red color, indicating the presence of blood. There are numerous small, dark purple-stained cells scattered throughout the field. Some of these cells are atypical, showing irregular shapes and sizes, and some have large, dark nuclei. There are also some larger, more complex structures that appear to be clusters of cells or debris. The overall appearance is consistent with a hemorrhagic background containing atypical cells.

Hemorrhagic background with atypical cells.

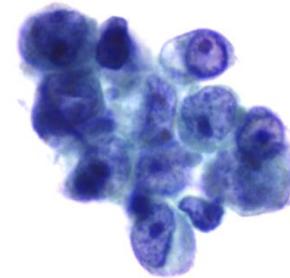
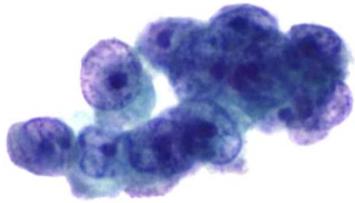
# Cytology

Sometimes isolated

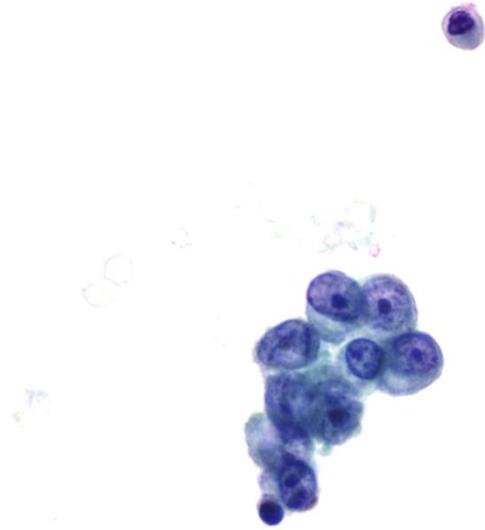


# Cytology

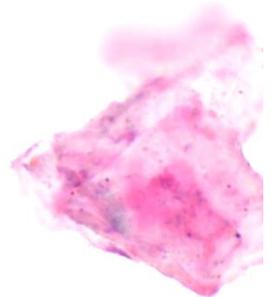
Or in some clusters



# Cytology



Slightly irregular and ovoid nuclei with fine granular chromatin and prominent nucleoli

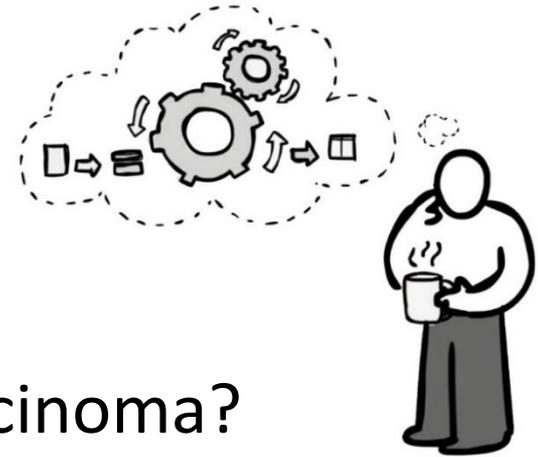


# Cytology

- Not features of cervical intra-epithelial lesion or squamous lesion.

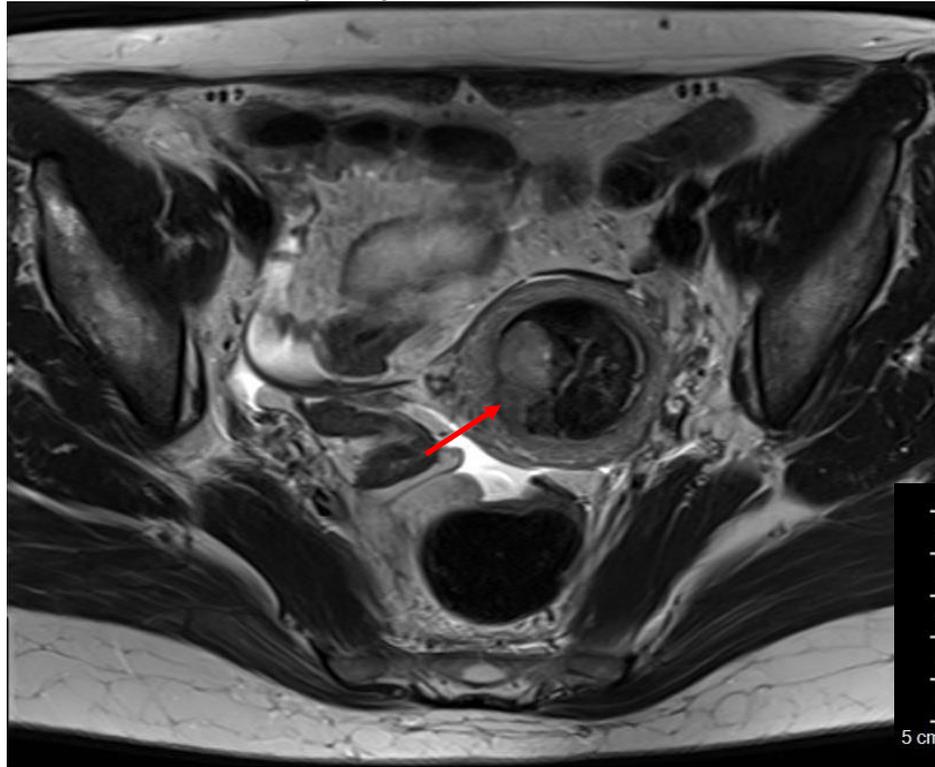
## **Possible differential diagnosis :**

- Endometrioid adenocarcinoma?
- Adenocarcinoma, NOS?
- Low-grade serous or clear cell carcinoma?
- Low grade Mixed Müllerian Tumor (MMT) (epithelial component)?
- Other hypothesis ?



# Clinical findings

- **Hysteroscopy** : intraluminal uterin bulky hemorrhagic necrotic mass of about 10 cm delivered by the cervix.
- **MRI** : polypoid endoluminal uterin mass without infiltration into the endometrium. No lymph node metastasis.

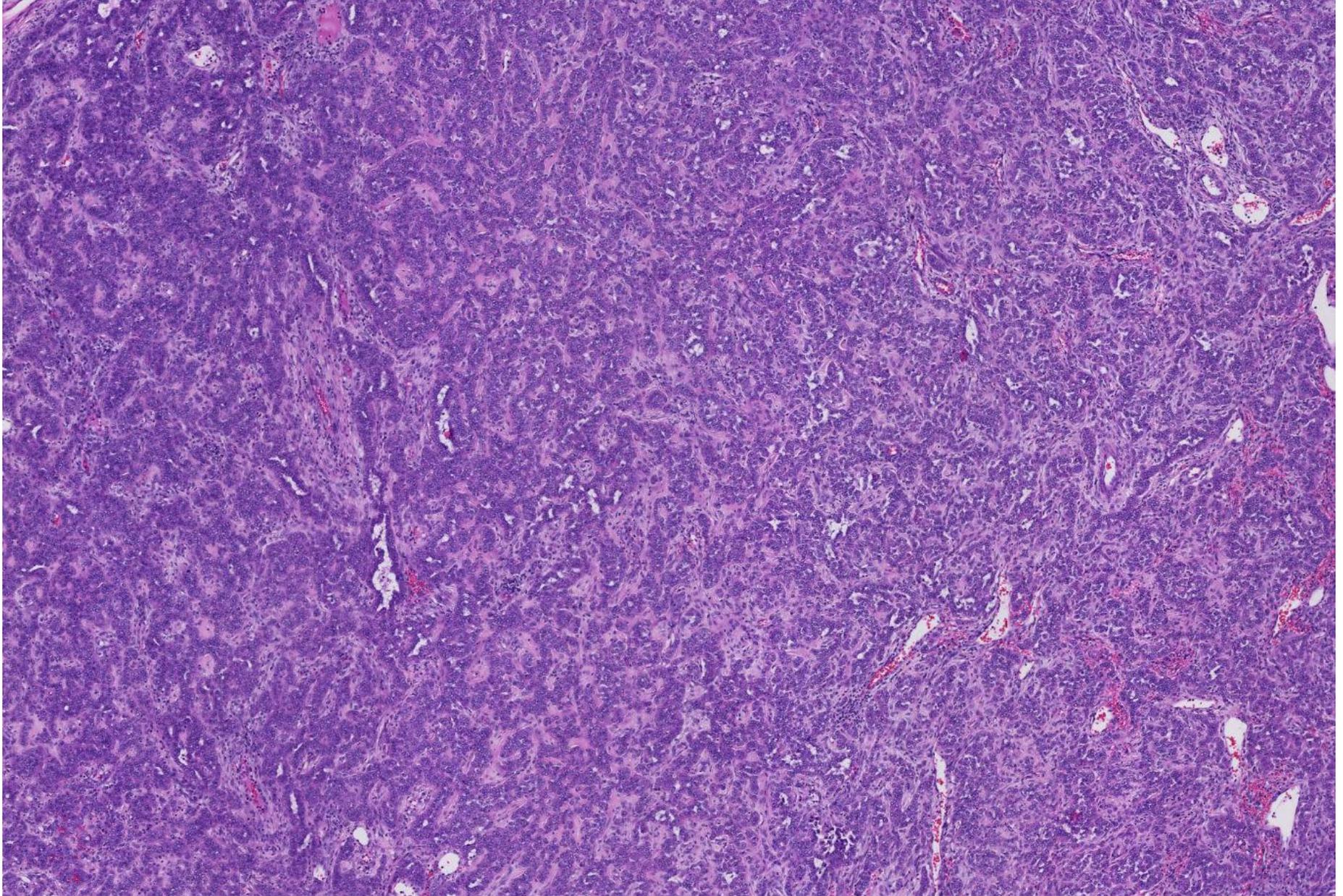


MRI



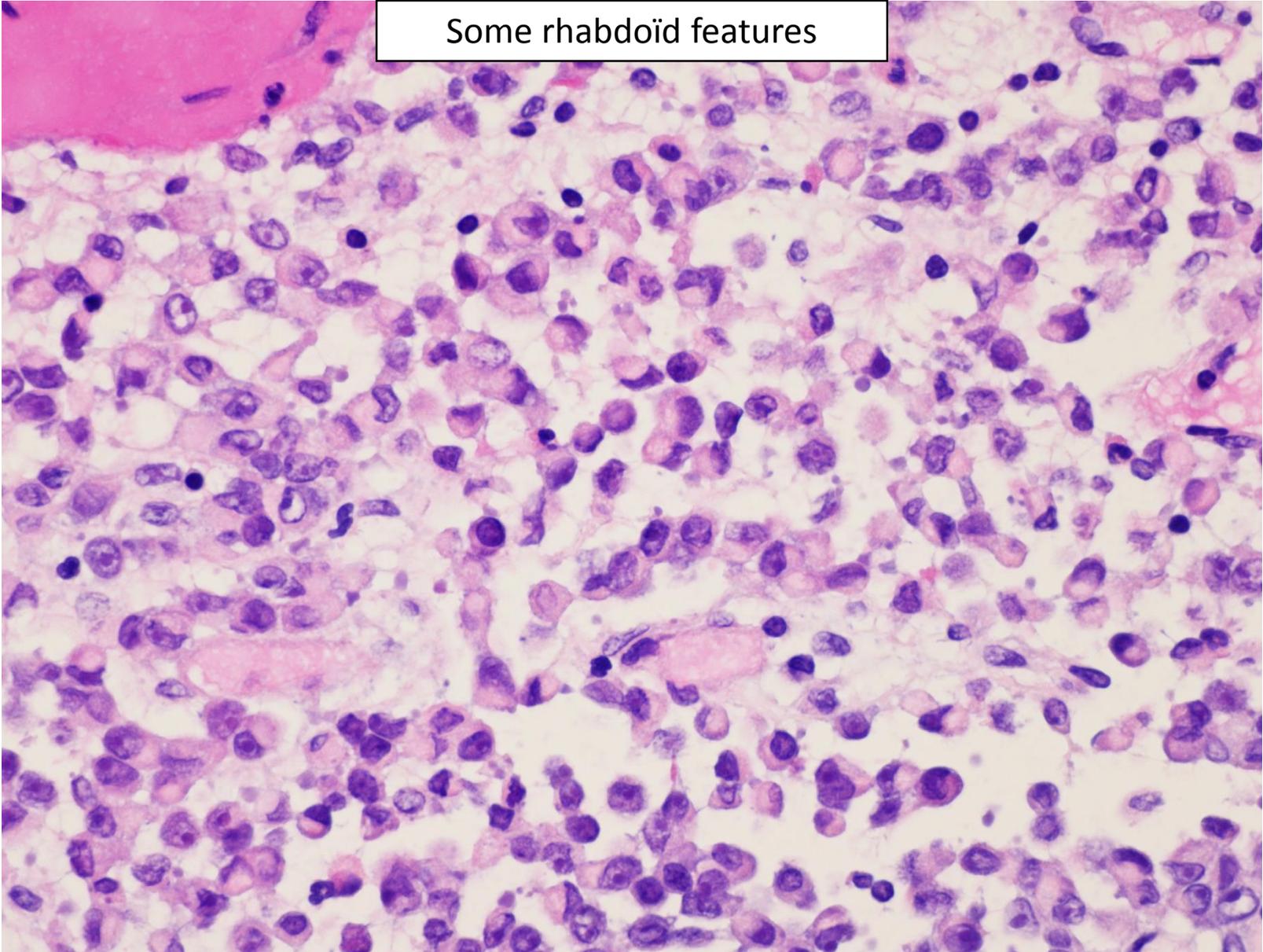
Resection of the mass by hysteroscopy (delivered fragmented)

# Histology

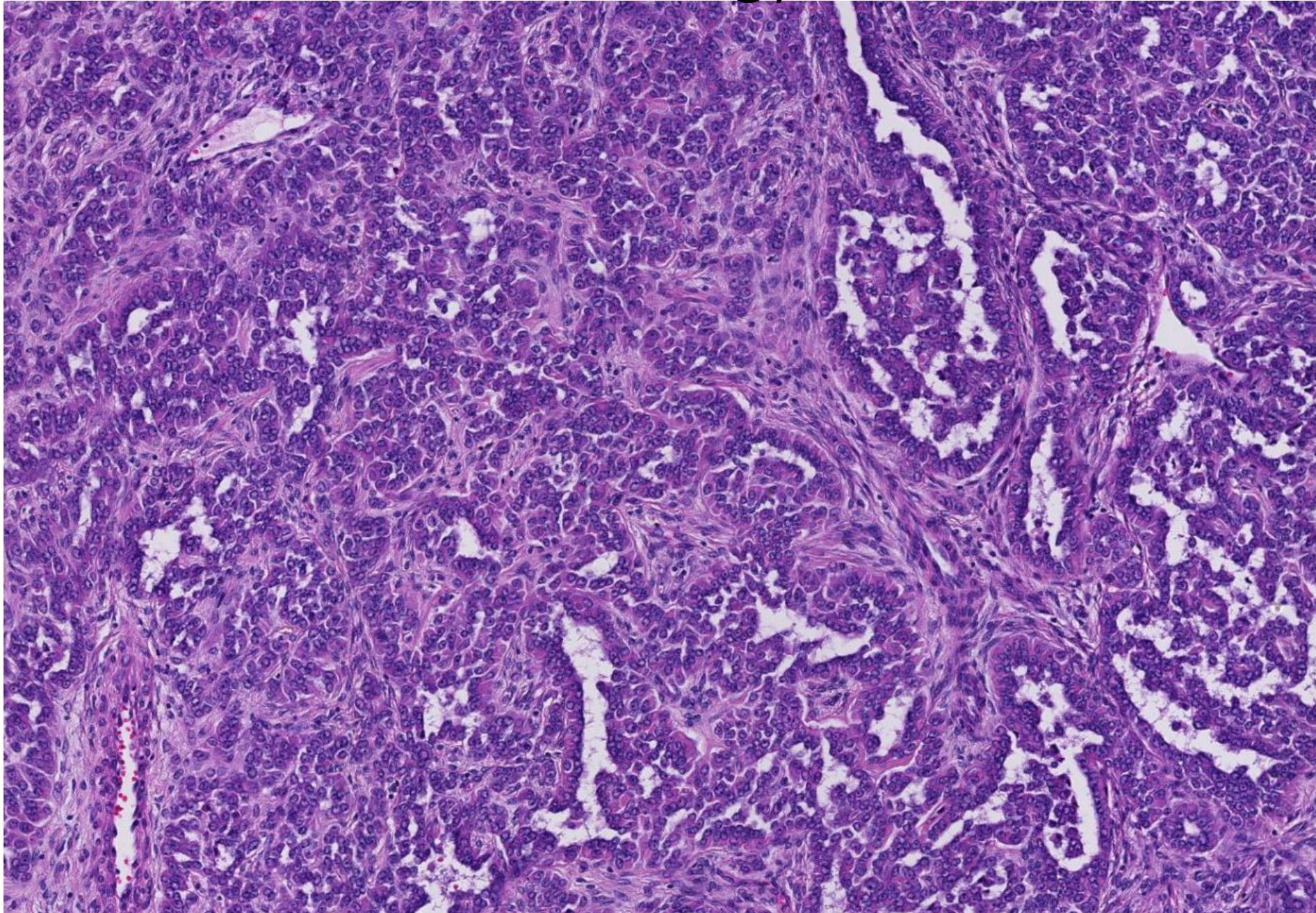


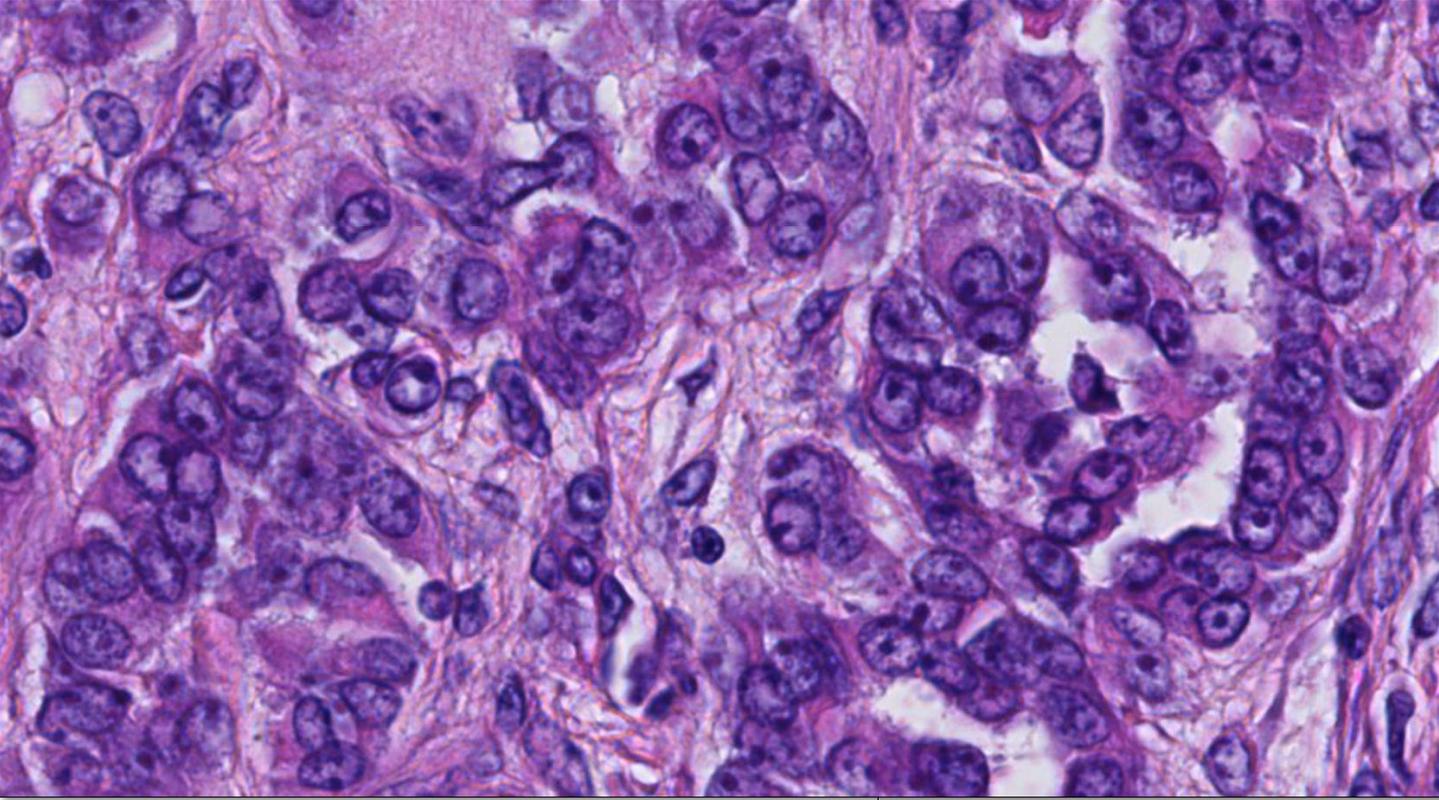
# Histology

Some rhabdoid features

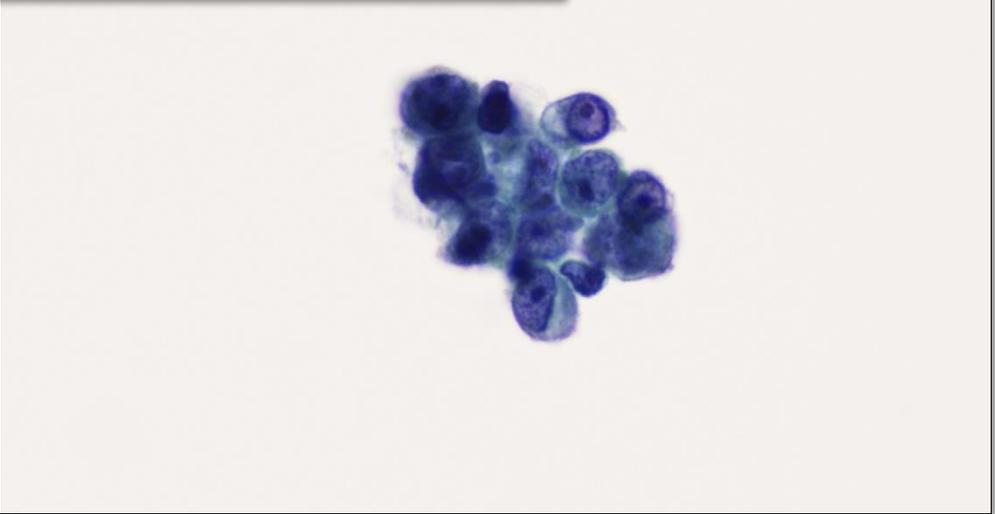


# Histology

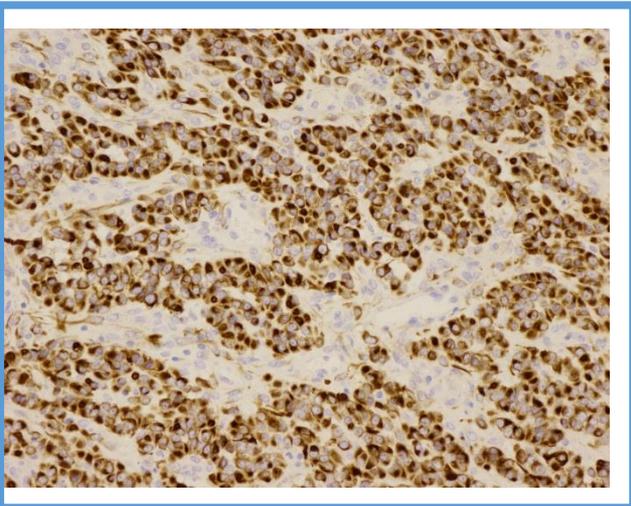




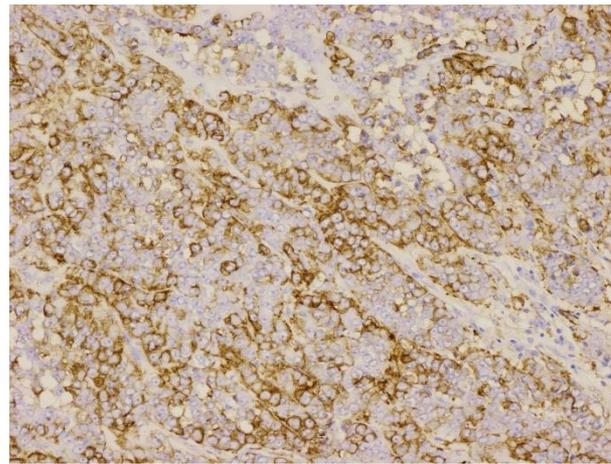
Same as cytology !!



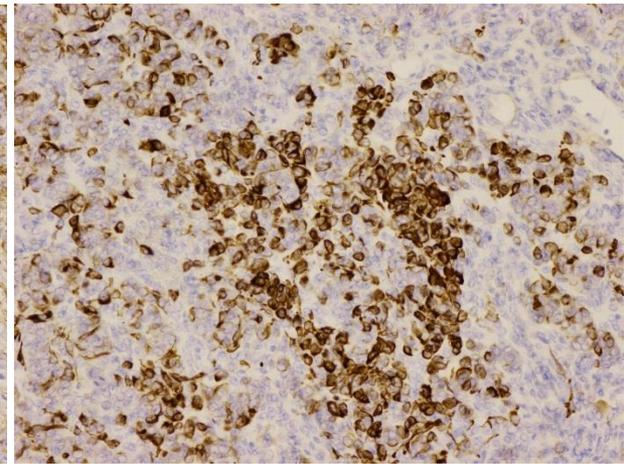
# Immunohistochemistry



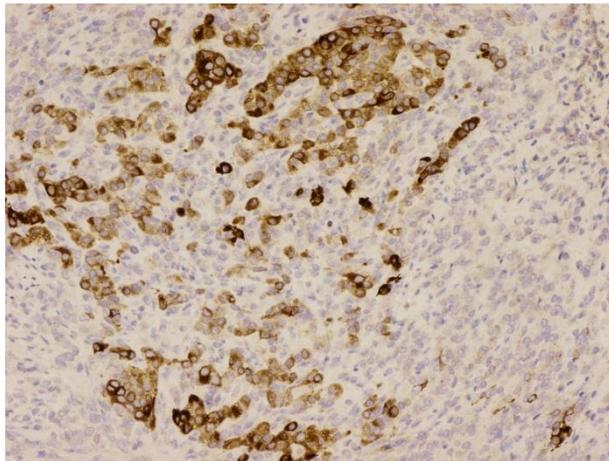
CKAE1/AE3



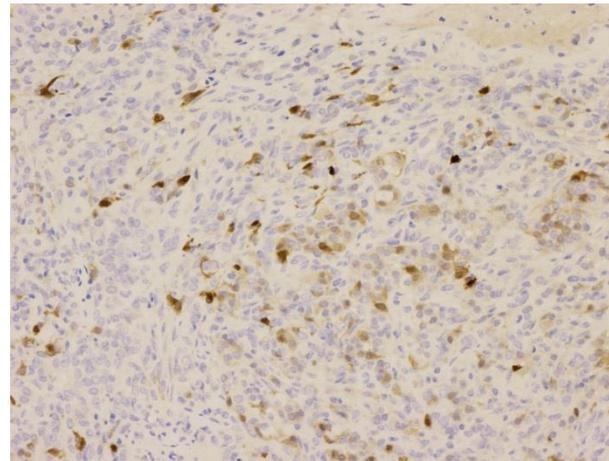
AML



Desmine



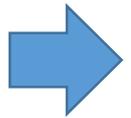
Inhibine



calrétinine

# Immunohistochemistry

- CKAE1/AE3 : + diffuse
- ER : diffuse
- CD56 : diffuse
- CD10 : focal
- Desmine, AML : focal
- MelanA, calrétinine, inhibine, WT1, CD99 : focal
- PAX8, CEA, CK7, CK20, napsinA : -



Final diagnosis?

# FINAL HISTOLOGICAL DIAGNOSIS:

## UTROSCT

- What is this??



- Uterin Tumor Resembling Ovarian Sex Cord Tumor

WHO 2014 : rare neoplasms that resemble ovarian sex cord tumors, without a component of recognizable endometrial stroma

# UTROSCT

- An uncommon uterine neoplasm of uncertain histogenesis
- Accounts for less than 0.5% of all uterine malignancies and 10% - 15% of mesenchymal uterine malignancies
- First reported by Clement and Scully in 1976 that classified these neoplasms into two groups:
  - Group I is endometrial stromal tumors with foci of sex cord differentiation (ETSCLE) < 50% (associated with recurrences and metastases)
  - Group II composed predominantly or exclusively by sex cord-like elements; this latest group continues to be classified as UTROSCTs

# UTROSCT

- Present mostly as intramural, less frequently submucosal, subserosal or polypoid / intracavitary masses
- Morphologically shows overlap with ovarian sex cord tumors with organization in sheets, cord, nests, trabeculae or tubules.
- Neoplastic cells are epithelioid with scant eosinophilic or clear cytoplasm and bland nuclei with minimal atypia and rare mitoses
- Coexpression of **epithelial** (KL1, CK AE1 / AE3, CAM5.2, EMA), **smooth muscle** (smooth muscle actin, desmin, h-caldesmon, smooth muscle myosin heavy chain, histone deacetylase-8), **sex cord markers** (WT1, calretinin, inhibin, CD99, MelanA, CD56, FOXL2, SF1) and **hormone receptors** (ER, PR, AR)
- Absence of JAZF1 / SUZ12 fusion (JAZF1-JJAZ1) and PFH1 gene rearrangements distinguish these neoplasms from endometrial stromal tumors

# UTROSCT

- Treated by surgical removal, based on age and parity of the patients (total hysterectomy with bilateral adnexectomy or conservative surgery with hysterectomy / mass resection)
- Most tumor have benign behavior without infiltrative properties but should be considered to have uncertain malignant potential

(recent study with 39 months follow-up of 34 patients, reports (23.5%) of extrauterine metastasis (pelvic, abdominal, peritoneum, ovary, lymph nodes, bone, liver and lung) and 8.8% of death.

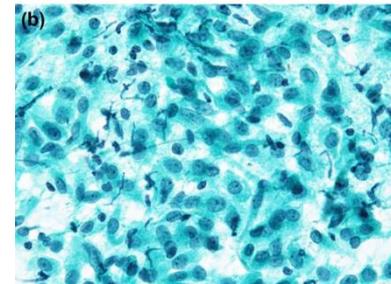
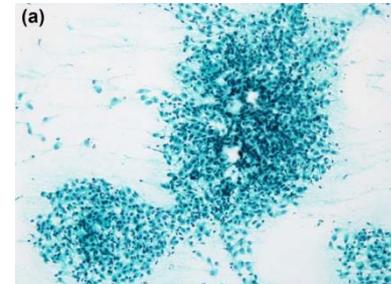
*Moore, 2017. "Uterine Tumour Resembling Ovarian Sex Cord Tumour: First Report of a Large Series with Follow-up." Histopathology )*

# First case of UTROSCT on Pap smears!

- Only two cases of cytological description (touch imprint) in the littérature :

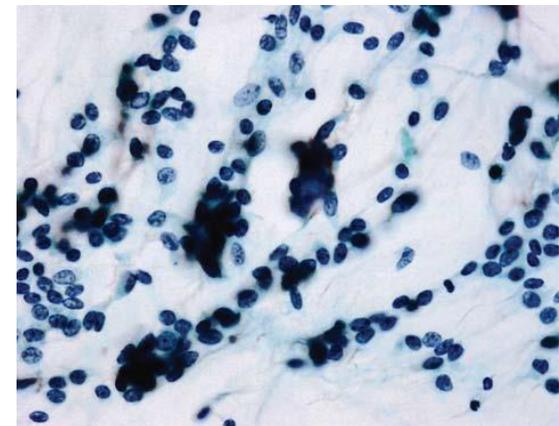
- one case of lung metastases from a UTROSCT

Kondo, 2018. "Lung Metastases of a Uterine Tumor Resembling Ovarian Sex-Cord Tumor: Report of a Rare Case." *Diagnostic Cytopathology*



- one case of low grade endometrial stromal sarcoma with sex cord-like differentiation

Nishikimi, 2010. "Cytologic Findings of Low Grade Endometrial Stromal Sarcoma with Sex Cord-like Differentiation: A Case Report." *Acta Cytologica*



# Conclusion

- It is the first case of UTROSCT in LBC Pap smears
- features may be confused with more common tumors with worse prognosis (endocervical adenocarcinoma, endometrioid adenocarcinoma, LG endometrial stromal sarcoma, epithelioid leiomyosarcoma)
- be aware to include UTROSCT in the differential diagnosis in cases that :
  - contain isolated or little cluster of mildly atypical ovoid cell with one prominent nucleoli;
  - in women presenting uterine mass protruding in the vaginal cavity (DD with MMMT).

# References

- Pathology outlines [www.pathologyoutlines.com](http://www.pathologyoutlines.com)
- Kondo, Yasuto, Shinobu Sakaguchi, Masashi Mikubo, Masahito Naito, Kazu Shiomi, Makoto Ohbu, and Yukitoshi Satoh. 2018. "Lung Metastases of a Uterine Tumor Resembling Ovarian Sex-Cord Tumor: Report of a Rare Case." *Diagnostic Cytopathology* 46 (1): 88–91. <https://doi.org/10.1002/dc.23804>.
- Moore, Michelle, and W. Glenn McCluggage. 2017. "Uterine Tumour Resembling Ovarian Sex Cord Tumour: First Report of a Large Series with Follow-up." *Histopathology* 71 (5): 751–59. <https://doi.org/10.1111/his.13296>.
- Nishikimi, Kyoko, Nobuya Habuka, Yuko Okazima, Shinichi Tate, Yuichiro Nagai, Masayuki Nakano, Reiko Okawa, and Makio Shozu. 2010. "Cytologic Findings of Low Grade Endometrial Stromal Sarcoma with Sex Cord-like Differentiation: A Case Report." *Acta Cytologica* 54 (1): 85–88. <https://doi.org/10.1159/000324975>.
- Leval, Laurence de, Gkeok Stzuan Diana Lim, David Waltregny, and Esther Oliva. 2010. "Diverse Phenotypic Profile of Uterine Tumors Resembling Ovarian Sex Cord Tumors: An Immunohistochemical Study of 12 Cases." *The American Journal of Surgical Pathology* 34 (12): 1749–61. <https://doi.org/10.1097/PAS.0b013e3181f8120c>.