

Overwhelmed by heterologous elements: Hidden identity of ovarian tumor.

A 21 year female presented with pelvic pain. CT Abdomen shows left ovarian mass measuring 40x38x35mm.

Conservative fertility sparing surgery i.e., left partial oophorectomy was performed and sent for histopathological examination.

On gross examination of ovarian mass, capsule was smooth without any surface deposits. Cut surface is grey white, firm to hard with calcified areas. Microscopy revealed encapsulated ovarian tumor comprising of predominantly calcifications, ossification, hyalinization admixed with tumor cells arranged in lobules, trabeculae and cords. Tumor cells show epithelioid morphology with abundant eosinophilic cytoplasm.

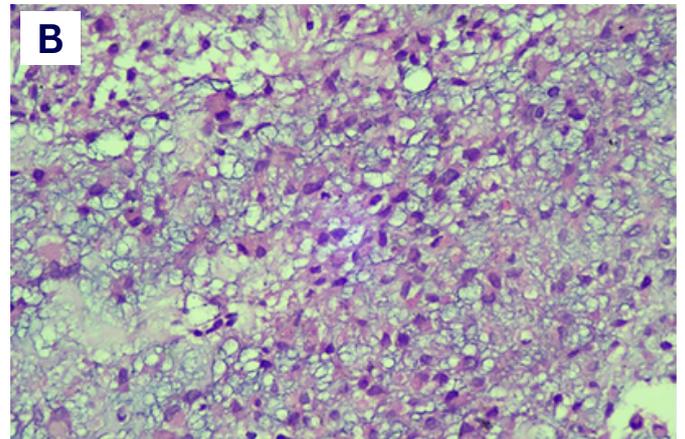
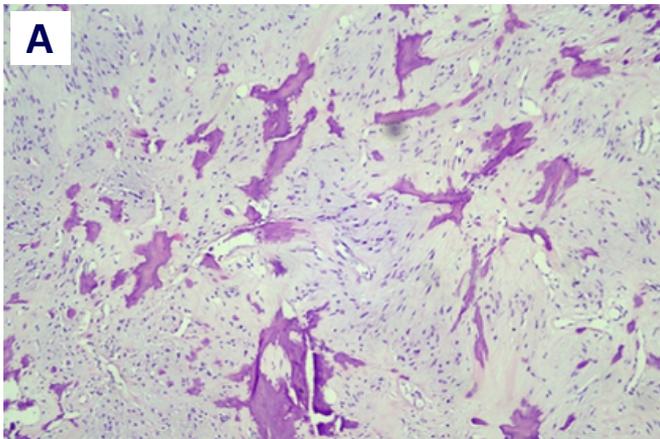


Fig1. (A) Low power view showed predominantly calcifications admixed with tumor cells (H&E, 10x). (B) High power view showed few cellular areas composed of tumor cells arranged in sheets, trabeculae and having moderate to abundant eosinophilic cytoplasm. (H&E, 40x)

Based on the morphological features, the differentials included were:

1. Teratoma
2. Mesenchymal tumor of ovary
3. Sex cord stromal tumor.

Case discussed with clinician regarding detailed case history. Patient had secondary amenorrhea and virilization symptoms like excess facial and body hair and presented to clinic with severe lower abdominal pain.

Immunohistochemistry was suggested for definite diagnosis.

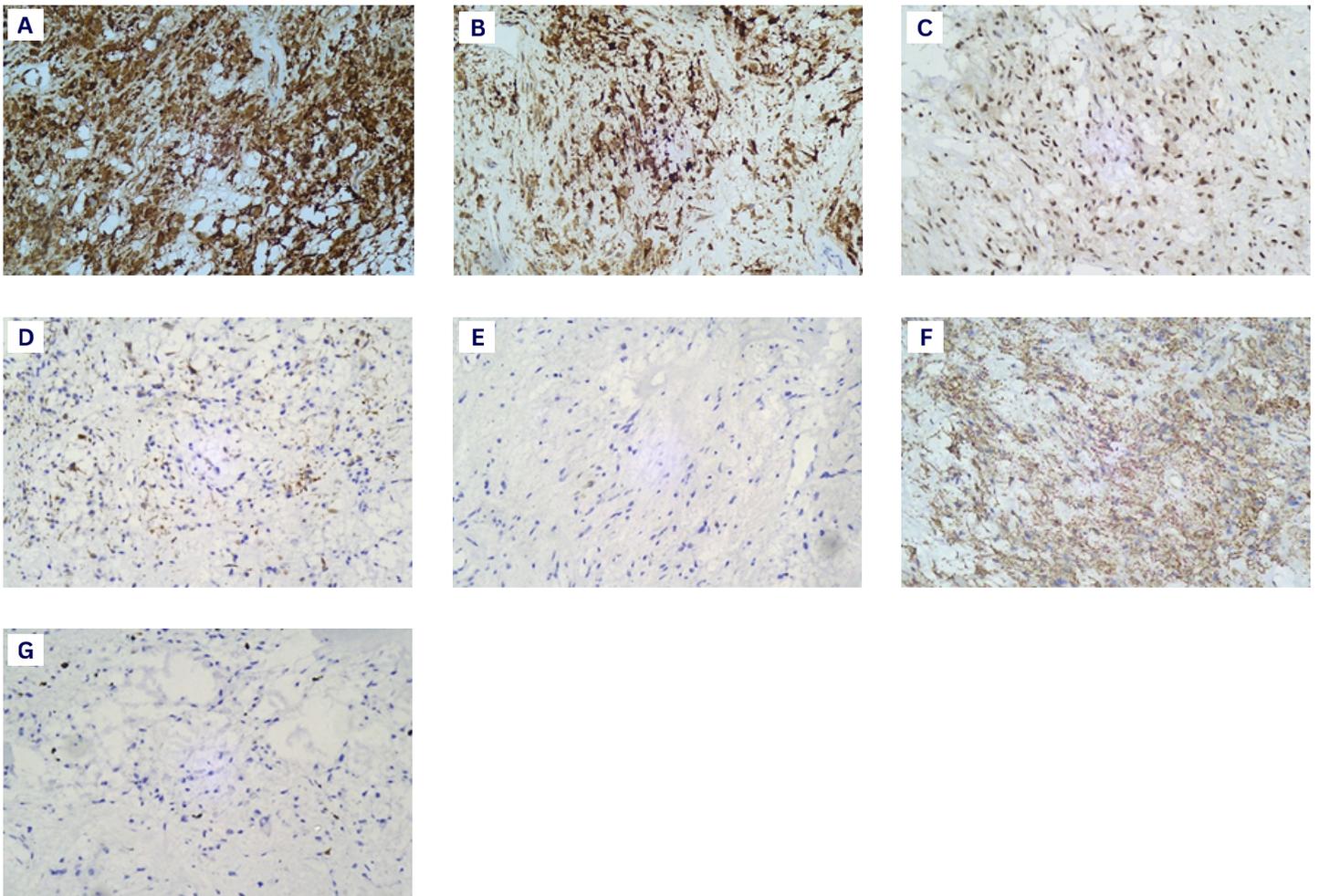


FIG2. Immunohistochemistry: (A) Inhibin (B) Calretenin (C) SF-1 (D) Pancytokeratin (E) S-100 (F) SMA (G) Mib-1 (IHC, 10x)

Tumor cells showed positivity for Inhibin, calretinin and SF-1 confirming sex-cord stromal origin. Pan CK is focal positive, S-100 and SMA immunostains were negative, ruling out mesenchymal origin. Mib-1 is low, 05%

Final diagnosis: Sertoli-Leydig cell tumor with Extensive heterologous elements

KEY FEATURES:

- Sertoli-leydig cell tumors are Rare, <0.5% of all ovarian neoplasms and 1-2% of pediatric ovarian cancers.
- Most Sertoli-Leydig cell tumors are seen in young women. Rare after menopause.
- Nearly 22% cases associated with heterologous elements.
- Half of the cases are associated with androgen excess manifested as defeminization and later masculinization.
- Mutations in DICER1 are associated with younger age at diagnosis, higher grade, heterologous elements and retiform pattern.
- FOXL2 Mutations are associated with postmenopausal patients, moderately/ poorly differentiated morphology without retiform and heterologous elements.
- Conservative fertility sparing surgery is the treatment modality in young patients.
- Platinum based adjuvant chemotherapy is beneficial.
- Genetic counselling and germline testing for DICER 1 mutation recommended.
- Tumors with heterologous elements, retiform pattern and tumor rupture are associated with poorer prognosis.

TAKE AWAY MESSAGE:

The presence of such extensive calcifications overshadowing the sparse Sertoli-Leydig cell component posed significant diagnostic dilemma. Immunohistochemistry and clinical presentation played crucial role in supporting the diagnosis. This case highlights the importance of high index of suspicion and employing multimodal diagnostic strategies. Distinguishing from other ovarian tumors helps in personalized treatment planning and improving long term survival outcomes.