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# A Fortuitous Case of Sex Cord Stromal Tumor - Presenting as Metastasis to Periumbilical Region: Cytohistopath Correlation and Review of Literature

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**Abstract:** Granulosa cell tumors are rare and their extraovarian location is further scarce in demography and various research studies. Origin from the mesenchyme of genital ridge predisposes to the extraovarian sites. Possibility of recurrence after initial surgery may occur. We present a case of extraovarian Granulosa cell tumor presenting as abdominal wall swelling. A high level of suspicion is required since the location may be increasingly rare and the treatment modality varies with follow-up periods. Cyto-histopathological correlation with accompanying Immunohistochemistry may aid in clinching the diagnosis.

## I. Introduction:

Granulosa Cell Tumours (GCT)belong to sex cord-gonadal stromal or non-epithelialgroup of tumourswhichconsist of granulosa cells,theca cells, and fibroblasts in varying proportions andcombinations.[1]

GCT are the most frequent among the malignant sex cord—stromal tumours of the ovary.[2] They can arise in extra-ovarian locations and may be derived from the mesenchyme of the genital ridge.[3]They can recur or metastasize manyyears after initial treatment and can rarely developat an extraovarian site, even in an oophorectomizedpatient.[4] Primary GCTs occurring at extraovarian sitesare rare, and having a primary tumour arising from theretroperitoneum is exceedingly rare.[4]Women who have had oophorectomy may have the potential to develop GCT later.[3]But primary extraovarian GCT is an extremely rare tumor.[5]

## II. Case report:

A 59 yrs old lady (Para 3, Living 3) with no known comorbidities, presented to the Surg Out Patient Dept with complaints of firm tender swellings in the periumbilical regionon the anterior abdominal wall, measuring 2x1x1.5 cm and 1.5x1 cm respectively. The swellings were seen approx. 3 months ago and were increasing in size steadily. The swellings were non reducible, cystic to firm and non-motile. Initial clinical impression was sebaceous cyst. Her Last Menstrual Period was 1 month back; her menses being irregular since last 6 months. On examination, pallor was mild, no icterus or any lymphadenopathy. Her pulse was 82 beats per minute and Blood pressure was 122/80 mmHg. Her investigations were as follows in Table 1:

Table 1.

| Sl No | Investigations | Values |
|---|---|---|
| 1. | Hb (gm/dl) | 10.4 |
| 2. | TLC | 9100 |
| 3. | DLC (P, L, M, E) | 75/21/2/2 |
| 4. | Platelets (/cmm) | 158,000 |
| 5. | PT (sec) | 16 (C -13 s) |
| 6. | INR | 1.4 |
| 7. | Blood sugar random (mg/dl) | 104 |
| 8. | S Urea (mg/dl) | 32 |
| 9. | S creatinine (mg/dl) | 0.9 |
| 10. | S bilirubin/ Direct (mg/dl) | 1.2/0.3 |
| 11. | SGOT/ SGPT | 46/28 |
| 12. | S Sodium (mEq/L) | 134 |
| 13. | S Potassium (mEq/L) | 5.0 |
| 14. | S LDH (U/L) | 171 |
| 15. | S amylase (U/L) | 33 |
| 16. | S lipase (U/L) | 144 |
| 17. | CA 125 | 21 (0-35 kU/L) |
| 18. | CA 19.9 | 6.1 (0-37 U/ml) |
| 19. | CEA | 2.4 (<3 µg/L) |

The swelling was subjected to Fine Needle Aspiration Cytology (FNAC) which yielded 0.2 ml of blood mixed fluid.The aspirates were cellular and showed monolayered sheets and clusters of oval to elongated cells, bland nuclei and characteristic coffee bean nuclei. FNAC was suggestive of Benign mesenchymal tumor (Figure 1,2).

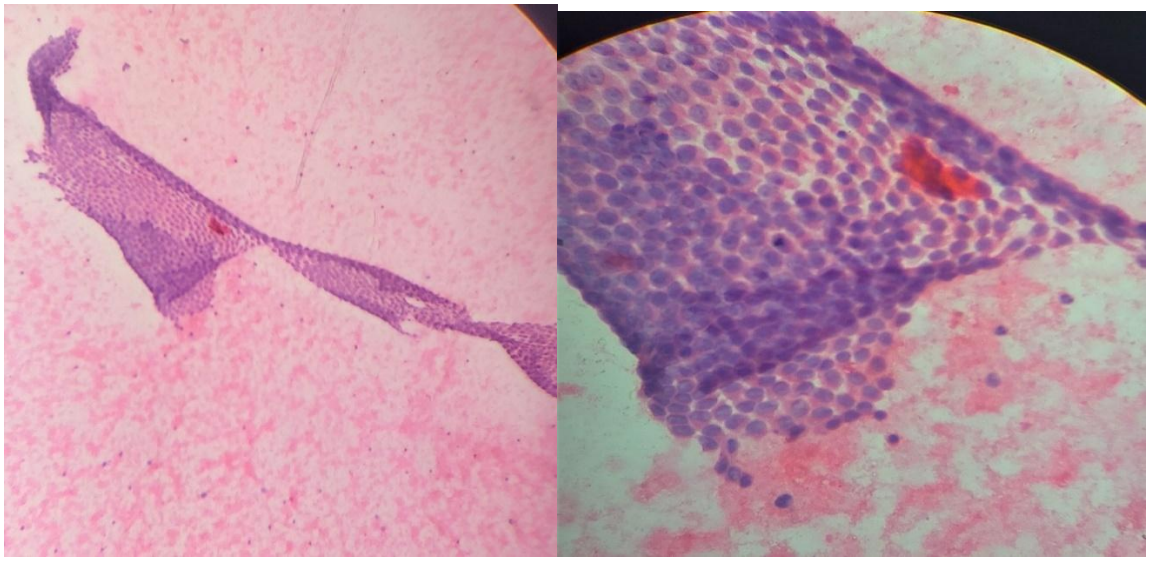

Figure 1,2. Fine needle aspiration cytology (LG stain): monolayered sheets of elongated cells with scanty cytoplasm

The nodule was excised and submitted for histopathology exam (HPE).Grossly the tumourmeasured 1.5x0.9x0.9 ccm. It was partly cystic and partially solid with areas of intracystic haemorrhage. Haematoxylin& Eosin (H&E) stained sections from the Formalin Fixed Paraffin Embedded (FFPE) blocks showed diffuse sheets, nodules and cords of atypical cells (Figure 3). These cells were round to oval, had high NC ratio, hyperchromatic to open chromatin, irregular nuclei, nuclear grooves with prominent nucleoli. Many lacunar spaceswere also noted. Mitotic figures were increased (2-5/10 HPF) with a few atypical mitotic figures also noted. Large areas of haemorrhage with hemosiderin pigments were seen. No epithelial lining or keratin debris were seen.

Immunohistochemistry revealed the tumourcells to be Inhibin, Calretinin, WT1 positive, FOXL2 positivewith focal positivity for S100 (Figure 3). Post Immunohistochemistry, the diagnosis was revised to Metastases from Sex cord stromal tumour – Granulosa cell tumour.

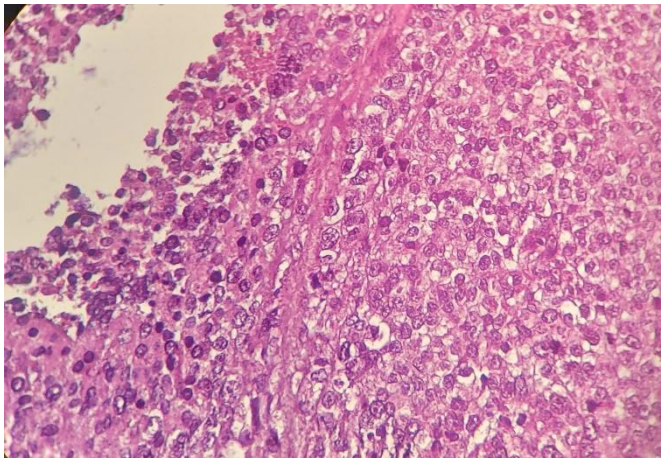

Figure 3 (a). FFPE Sec (H&E stain): sheets and nests of elongated to spindle cells with coffee bean nuclei

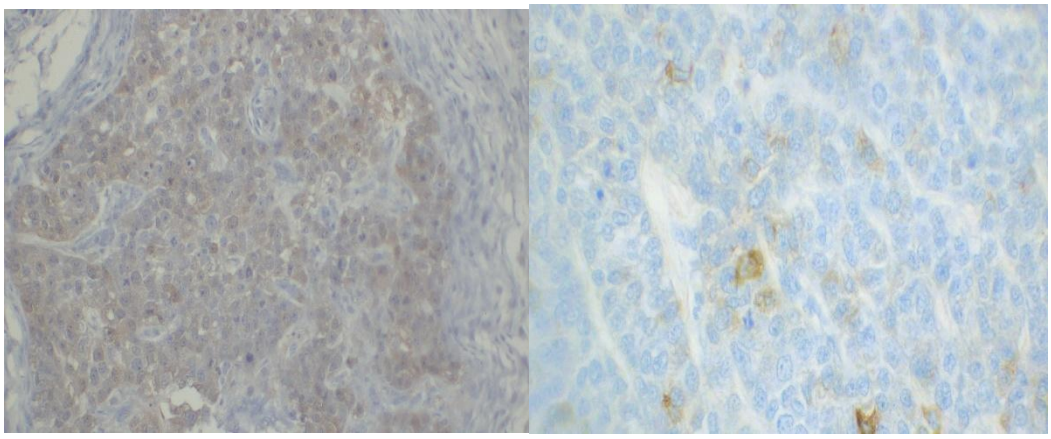

Figure 3 (b). Calretinin positive Figure 3 (c). Inhibin positive

The patient was operated and is presently asymptomatic after 1 yr of surgery. No recurrence was detected on follow-up.

## III. Discussion:

Granulosa cell tumours (GCT) of the ovary are malignant sex cord-stromal tumours. They form 2% of all ovarian tumors.[2] These tumours can be classified as adult or juvenile types according to the age of presentation, clinical and histological features. The adult type of GCTs are commonly encountered and are seen in the perimenopausal or early menopausal period (median age 50-54 yrs).[4] Patients with GCT need longer follow-up periods with history, clinical examination and assays of tumours markers as relapses to the tune of 17% are known which can occur even after 10 yrs of initial diagnosis.[4]Among the sites, pelvis is most common for occurrence of GCTs. Though rarely they have been found to develop in extra-ovarian sites like retroperitoneum, broad ligament, fallopian tubes[6], mesentery, liver, adrenals[7]and omentum.[8]They tend to recur or metastasise many years after initial management. GCTs can develop at extra-ovarian sites in rare scenarios. Any possibility of metastasis has to be eradicated before the diagnosis of extraovarian GCT. Extra-ovarian GCT is an uncommon tumour with only occasional cases reported in English literature and rarely found in the Indian subcontinent.[4,9] The possibility of metastases needs exclusion before diagnosing a case as Extraovarian GCT.

It is postulated that the tumour cells originate from ectopic gonadal stromal tissue from the mesonephros. [4] A dual derivation from boththe mesonephros and coelomic epithelium hasbeen suggested by other authors. The mesonephros seems to beessential for the development of the sex cord.This might propose the growth of GCTs in theretroperitoneum, the broad ligament, or the adrenal,all of which differentiate in close proximity to themesonephros and the mesonephric duct[7].

A search for extraovarian GCT was conducted in Pubmed central, Google Scholar and Scopus which revealed 11 cases after using the keywords extraovarian, case report or autopsy report and eliminating the review or repetitive cases. However; no case of extraovarian GCT in the anterior abdominal wall was found.
The aim of this study was topresent an unusual case of Extraovarian presentation of granulosa cell tumor. The inherent dilemma faced while reporting a sex cord stromal tumor, correlation with clinical features and a high index of suspicion are required to diagnose Granulosa cell tumor. Cyto-histopathological correlation assumes higher significance as we have seen in this case where cytology clearly showed nuclear grooves (coffee bean nuclei) characteristic of the adult variant of Granulosa cell tumor.

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