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Pathology

Gynandroblastoma: A Rare Case Presentation

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Abstract **Case Report**

Gynandroblastoma is a rare mixed sex cord stromal tumor of ovary. These tumors contain both granulosa cell component and Sertoli-leydig cell component. Very few cases of Gynandroblastomas are reported in literature. Reported cases are accompanied by estrogenic, androgenic or no harmone effects. This is a case of Gynandroblastoma reported in 18 year old female with history of pain abdomen, spotting per vaginum, significant weight loss since 3 months. The tumor diagnosis is based on histopathological features.

Keywords: Gynandroblastoma, Granulosa Cell Tumor, Sertoli Cells, Leydig Cells.

INTRODUCTION

Gynandroblastoma is an extremely rare mixed sex cord stromal tumors of ovary. Robert Meyer first described this tumor in 1930. This tumor histologically manifests as combination of granulosa cell and Sertoli-Leydig cell components. The granulosa cell component is usually adult type. Gynandroblastoma with juvenile type of granulosa cell tumor is much rarer [1]. Gynandroblastoma most commonly present in age group less than 30 years.

However, very few cases have been reported in post menopausal women [2]. It is very difficult to characterise biological behaviour of ovarian gynandroblastoma due to its low incidence. Based on the reported cases in the literature, it appears to have a benign course and most of the patients present with stage I tumors [2].

Initial work-up starts with evaluation of tumor markers inhibin, estradiol, testosterone, Alpha fetoprotein and pelvic ultrasound. Cross sectional imaging with CT or MRI is typically required [3]. DICER1 mutations are reported in few cases of gynandroblastoma [3].

CASE HISTORY

A 18 year old unmarried female patient came to gynecology OPD with complaint of amenorrhea, weight loss and spotting per vagina since 3months and pain abdomen since one month. Initial biochemical work for tumor markers was done and values are interpreted as follows:
βhcg- 1.20mIU/L
Inhibin A- 40.7pg/ml
Inhibin B-379pg/ml
CA125-145.7µ/ml
Alpha fetoprotein- 2.1ng/ml
Carcinoembryonic antigen-1.9ng /ml

Imaging studies (MRI) show large well defined T2 hyperintense mass with cystic spaces in the right adenexa. The imaging features are consistent with dysgerminoma of right ovary. Patient has undergone right salphingo oophorectomy and specimen was sentto department of pathology. The specimen was fixed in 10% buffered formalin. Gross description of tumor is single globular mass measuring 14x13x10 cm. External surface is grey white to grey brown. Cut surface is grey white with few grey brown areas with variegated appearance showing both solid and cystic areas. Normal ovarian parenchyma is not visualized. Tube measuring 6cm identified. No abnormality is detected in external surface of fallopian tube. Lumen identified in cut surface of tube. Multiple sections were taken from formalin fixed and paraffin embedded blocks and stained with hematoxylin and eosin stain.

Figure 1: External surface showing engorged veins

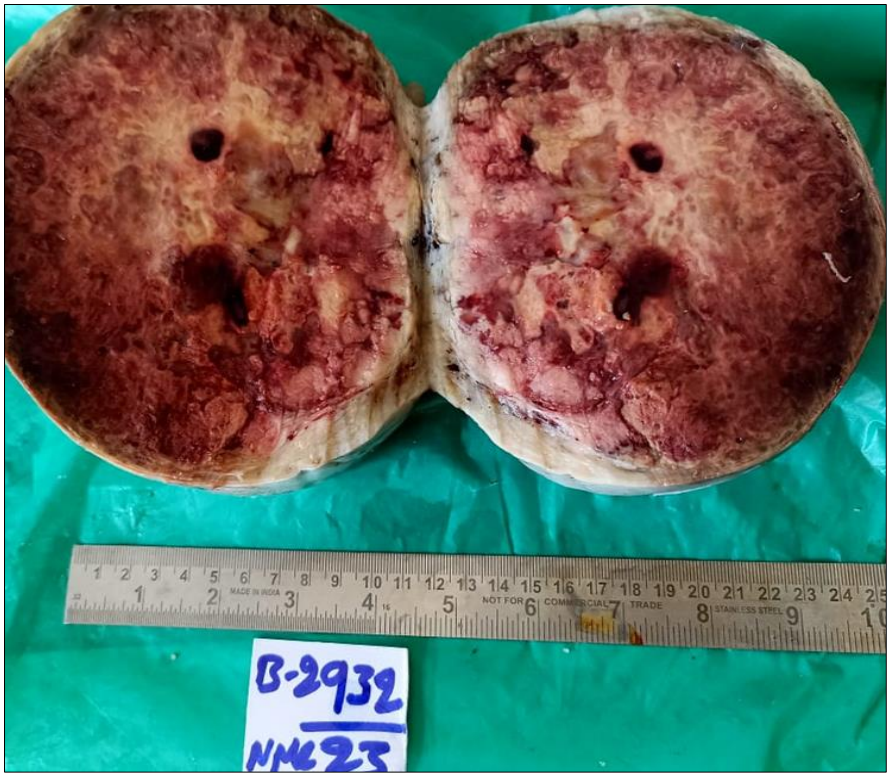

Figure 2: Cut surface showing heterogenous organs with both solid and cystic areas

Multiple sections examined under light microscope reveal tumor consisting of round polygonal cells in glandular and microfollicular pattern forming occasional Call-Exner bodies. The cells have vesiculated nuclei and some of them show coffee bean nuclei. The stromal cells are bigger with clear cytoplasm. They are seen in clusters. Also seen are oval to spindle cells forming tubules, cords, and trabecular pattern. The cells have vesicular nuclei with prominent nucleoli and clear cytoplasm. Amidst there are large cells with abundant granular cytoplasm with prominent nuclei. Dense foci of necrosis, hemorrhage and cyst formation are seen.

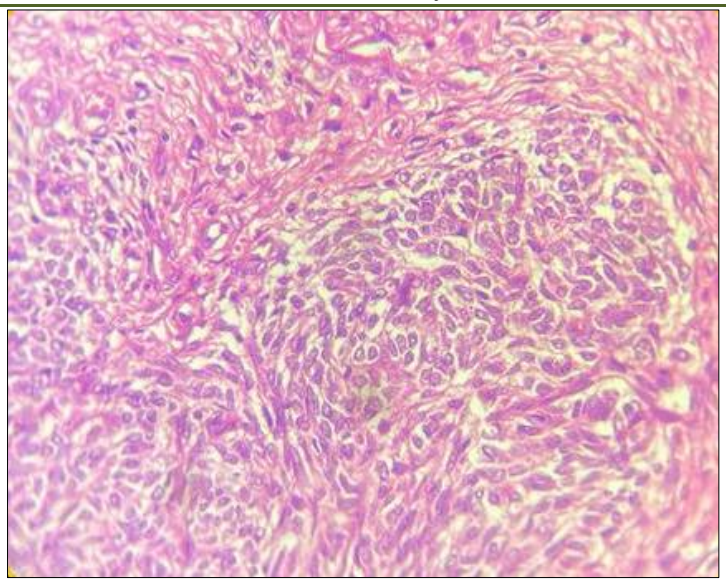

Figure 3: The cells have vesiculated nuclei and some of them show coffee bean nuclei. (H&E, X 400)

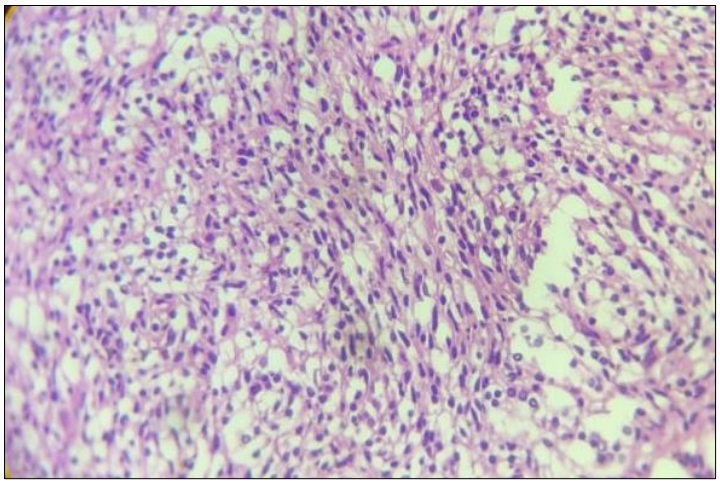

Figure 4: Oval to spindle cells forming tubules, cords and trabecular pattern. The cell have vescicular nuclei with prominent nucleoli and clear cytoplasm (H&E, X 400)

DISCUSSION

Ovarian gynandroblastoma (GAB) is an extremely rare sex cord-stromal tumor showing morphological evidence of both female (granulosa cell tumor) and male (Sertoli–Leydig cell tumor (SLCT)) components [4].

Almost all the cases have been reported in children, adolescents, or women of reproductive age. In our case the age of presentation is 18 years.

The main clinical manifestations in order of frequency are amenorrhea; metrorrhagia; isolated pelvic mass syndrome [5]. In the present case, patient presented with amenorrhea, weight loss and spotting per vagina since 3 months.

In the case study done by Hawang S *et al*., increased CA125 levels is seen in 65 year old woman with gynandroblastoma [4].

In the present case increased CA125 levels is noted.

The granulosa cells in gynandroblastoma usually show the characteristics of adult-type granulosa cells. However, in extremely rare occasions, juvenile-type granulosa cells constitute the granulosa cell component of gynandroblastoma [6].

In the study done by Takeda *et al*. in 15 year old prepubertal girl, the granulosa cell component is juvenile granulosacell type [7]. In the present study, the granulosa cell component is adult type.

Gynandroblastomas has to be differentiated from other mixed sex cord stromal tumors. Sertoli Leydig cell component in gynandroblastoma shows DICER 1 mutations in most of the cases [8].

CONCLUSION

Gynandroblastoma is a rare ovarian tumor that can be seen in adolescent girls as well as in post menopausal women. Clinical signs of virilization and harmonal imbalances may or may not be present posing diagnostic challenges. Most of the cases of gynandroblastoma presents with mass per abdomen and pain abdomen.

Though CA125 levels are increased in few cases of gynandroblastomas, the role of tumor marker assays are not codified in the literature. DICER1 mutations are seen in few cases of gynandroblastoma. Hence CA125 assays and DICER1 mutation analysis can help in diagnosis. Of all the published cases of gynandroblastoma till date, there is no post operative recurrence of tumor.

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