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A RARE CASE REPOT: JUVENILE GRANULOSA CELL TUMOUR

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Conflicts of Interest: Nil

ABSTRACT:

Ovarian cancer is the third most common neoplasm of the female genital tract. Based on the cell type of origin, primary ovarian malignancies are classified into surface epithelium, germ cell, and sex cord tumors. Sex cord tumors account for 1% to 2% of ovarian malignancies. Juvenile Granulosa Cell Tumours (JGCTs) are clinically & histopathologically distinct from the GCTs. They are rarely encountered but mostly in youngsters. Surgery is the primary modality of treatment with chemotherapy being reserved for advanced or recurrent disease states. We herewith report an interesting case of JGCT in a young teenage girl.

Keywords: Ovarian sex cord stromal tumours, Juvenile granulosa cell tumour

Introduction

Granulosa Cell Tumours (GCTs) account for approximately 2-5% of all ovarian tumors and can be divided into adult (95%) and juvenile (5%) types based on histologic findings. Juvenile Granulosa Cell Tumors (JGCTs) are rare sex-cord stromal tumours occurring in the younger age groups. JGCT is clinically & histopathologically distinct from the GCTs. We here with report an interesting case of JGCT in a young girl.[1]

CASE REPORT

A 14 year old unmarried girl came with complaints of irregular cycles since 6months duration.

On clinical examination General condition good.

Vitals: Within Normal Limits (WNL).

Heart and lungs clinically normal

Secondary sexual characters well developed.

Abdominal examination A non-tender mass of 16 weeks size occupying the lower abdomen, firm in consistency, mobile from side to side & lower border of the mass could not be made out.

Vaginal examination not attempted

Per rectal examination

Vague fullness present in pouch of Douglas. Uterus could not be felt separately. Rectal mucosa free.

Investigations Baseline investigations: WNL.

Thyroid function tests& serum prolactin: normal.

Tumour markers Serum CA-125, CEA, AFP, β-hCG & LDH were WNL.

Ultrasound

Large 17×16 cm sized, well defined mass, predominantly cystic with increased vascularity is noted in the hypogastric region?arising from left ovary. No calcification or solid component noted. Uterus normal in size and echotexture. Right ovary visualised normal. Liver, spleen, gall bladder and kidneys were normal. No e/o of free fluid in peritoneal cavity.

Chest X-ray -Normal.

Management

Exploratory laparotomy.

Histopathology report:

Gross: ovarian cyst with attached fallopian tube measuring 17×16×5cms and fallopian tube measuring 5 cms in length. External surface well encapsulated with prominent vascular marking. On cutting open multiple cyst identified largest

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measuring 10cms in diameter filled with mucoid and gelatinous material alongwith solid area (grey brown to grey white in color) and hard in cosistency.

Microscopic features: tumor show round to ovoid tumor cells in varied pattern ranging from solid to nested, trabecular, follicular and watered silk patterns. There is no nuclear grooving seen. Mitosis is brisk focally with 3-4 mitotic figures per high power field and occasional atypical ones. Intermixed with this are areas showing thecal and leutinized cells. There are widespread areas of necrosis and haemorrhage.

Juvenile granulose cell tumour (well differentiated).

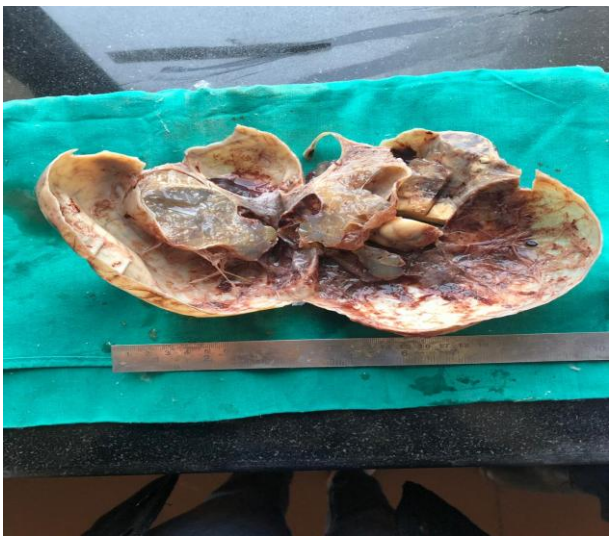

Figure 1: Macroscopic appearance

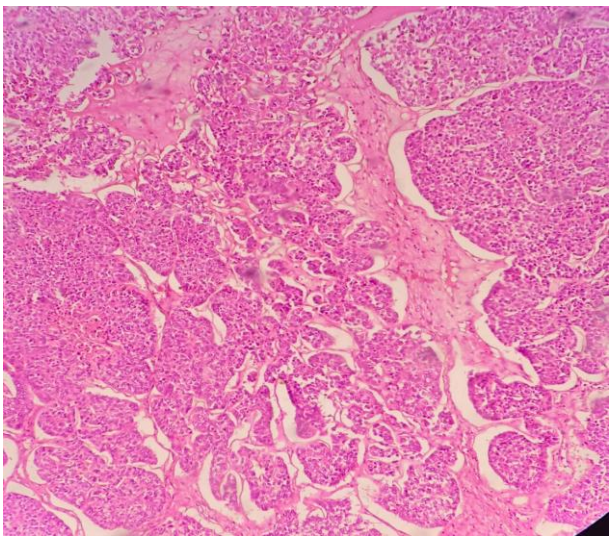

Figure 2: Histopathological appearance

DISCUSSION

Ovarian cancer is the third most common neoplasm of the female genital tract. Based on the cell type of origin, primary ovarian malignancies are classified into surface epithelium, germ cell and sex cord tumors. Sex cord tumors account for 1 to 2% of ovarian malignancies. They may contain granulosa cells, theca cells, sertoli cells, or fibroblasts of gonadal stromal origin. Approximately 70% of sex cord tumors are Granulosa Cell Tumours (GCT). Granulosa cell tumors have bimodal age distribution but the peak incidence is in postmenopausal period with median age of diagnosis around 50-55 years. [1] Juvenile Granulosa Cell Tumors (JGCTs) are rare sex-cord stromal tumours occurring in the younger age groups. GCTs were described for the first time in 1855 by Rokitansky. These tumors are malignancies with a relatively favorable prognosis. They are characterized by a prolonged natural history and a tendency to late recurrences. [2]

Imaging characteristics of adult and juvenile granulosa cell tumors are non-specific[3] and these tumors cannot be reliably distinguished from other ovarian neoplasms on imaging alone. On cross-sectional CT imaging and sonography, their appearance vary widely, but they often appear as a single large multiloculated cystic mass with solid components. They have multiple septations which can be thin, or thick and irregular. The adult form shows more variability with regards to the cystic component and can occasionally appear predominantly solid. Intratumoral haemorrhage, central areas of necrosis and fibrous degeneration can result in a heterogeneous solid appearance MR imaging is more distinctive. T1W MR images can demonstrate intracystic high signal suggesting characteristic intratumoral haemorrhage present in up to 71% of cases.

Patients with JGCT typically present at an early stage I and enjoy a favorable prognosis & five year survival of 95-100% if diagnosed at early stage. Recurrences are uncommon and typically occur within the first year. The incidence of this group of tumour is same throughout the world. Majority of JGCTs present as localized disease confined to the ovary, and usually behave in a benign manner despite having histopathological features of malignancy. However, those with more advanced-

stage disease (FIGO stages II, III, or IV) may experience an aggressive clinical course with a short remission to relapse or death. Both juvenile and adult type GCTs are almost always unilateral. As per the unilateral origin of the tumor with intact capsule & negative peritoneal cytology, our case was considered as stage 1A.[4]

JGCT is distinguished from AGCT by 1) more commonly occurring in young women (<30 years of age), 2) having follicles that are more irregular in shape and size, 3) lacking Call-Exner bodies, 4) having rounder nuclei that lack nuclear grooves, 5) typically having more abundant eosinophilic to vacuolated cytoplasm and 6) having follicles containing basophilic secretions. The histological report of our patient was consistent with the above mentioned JGCT features.

Though immunohistochemical staining for inhibin and smooth muscle Actin is positive in almost all cases, these tests could not be done in our patient due to specific problems like lack of reagents which could have given more distinctive information of JGCT and also help in monitoring for relapse in some cases. Vimentin positivity though not specific for the diagnosis of JGCT it is found to be helpful in distinguishing well differentiated GCT's from poorly differentiated ones. As per this the histopathology report in our case was given as "well differentiated juvenile granulosa cell tumour.[5]

CONCLUSIONS

Granulosa cell tumour is a rare neoplasm of different behaviour. In addition to its rarity in childhood, the presented case is of particular interest for having abdominal pain and distension only without any secondary sexual characteristics. Histopathological diagnosis requires a great deal of expertise in view of age, treatment modalities and prognosis as this tumour differs from other ovarian tumours. IHC markers may be prognostically helpful in this rare condition.

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