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Case Report

A rare encounter with juvenile granulosa cell tumor: diagnosis, management, and outcome

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ABSTRACT

Juvenile granulosa cell tumor (JGCT) is a rare ovarian sex cord-stromal tumor, accounting for less than 5% of all granulosa cell tumors and primarily affecting prepubescent girls and young women. Due to its rarity, clinical presentation, optimal management, and long-term outcomes remain areas of ongoing investigation. We report the case of a 19-year-old nulliparous female who presented with intermittent lower abdominal pain and bloating for eight months, with no menstrual irregularities or signs of virilization. Clinical examination revealed a firm, 20-week-sized pelvic mass, and imaging studies suggested a large multiseptated right abdominopelvic mass. Patient underwent fertility-sparing staging laparotomy, and histopathology confirmed JGCT with micro metastasis in the omentum (pT3aN0). Immunohistochemistry showed positivity for inhibin, PR, calretinin, and WT1. The patient underwent four cycles of adjuvant paclitaxel and carboplatin chemotherapy and has remained in remission for one year and under three-monthly surveillance. This case highlights the importance of early recognition, comprehensive surgical staging, and appropriate adjuvant therapy in managing JGCT. Given its potential for aggressive behavior and recurrence, long-term follow-up is essential. This report underscores the need for individualized treatment approaches in rare ovarian malignancies.

Keywords: Juvenile granulosa cell tumor, Ovarian sex cord-stromal tumor, Ovarian neoplasm, Fertility-sparing surgery, Adjuvant chemotherapy, Immunohistochemistry

INTRODUCTION

Sex cord-stromal tumors are rare ovarian neoplasms, comprising 3–5% of ovarian malignancies. They originate from primitive sex cords or stromal cells, including theca cells, fibroblasts## (GCTs) account for 70% of sex cord-stromal tumors, with an incidence of 0.4–1.7 per 100,000 women in developed countries. According to the WHO classification, GCTs are categorized into adult GCTs, which typically affect peri- and postmenopausal women, and juvenile GCTs, which are rare (5%), primarily occurring in adolescents and young women (<30 years old) but accounting for 67% of sex cord-stromal tumors in pediatric patients and 5–12% of ovarian neoplasms in childhood and adolescence. Clinical presentation includes abdominal pain/swelling and hormonal hypersecretion, manifesting as precocious puberty in prepubertal girls and virilization, hirsutism, acne, and menstrual irregularities in post pubertal patients, while management primarily involves surgery with or without fertility preservation, with adjuvant chemotherapy considered in select cases, though clinical data remain limited and guidelines on optimal management vary.[1]

CASE REPORT

A 19-year-old unmarried, nulliparous female presented to the gynaecology outpatient department with an eight-month history of intermittent lower abdominal pain accompanied by bloating she reported regular menstrual cycles with no history of menstrual irregularities or signs of hirsutism. On abdominal examination, a firm, 20-week-

sized pelvic mass with restricted mobility was palpated, and on rectal examination, the mass was separate from the uterus. Investigations included tumor markers CA-125, Alpha-fetoprotein (AFP), LDH, Beta-HCG, and CA 19.9, all within normal limits, transabdominal ultrasound revealing a large, multiseptated anechoic cyst measuring 18×9 cm, and contrast-enhanced magnetic resonance imaging (CEMRI) showing a large multiseptated right abdominopelvic mass, suggestive of a mucinous cystadenoma (Figure 1).

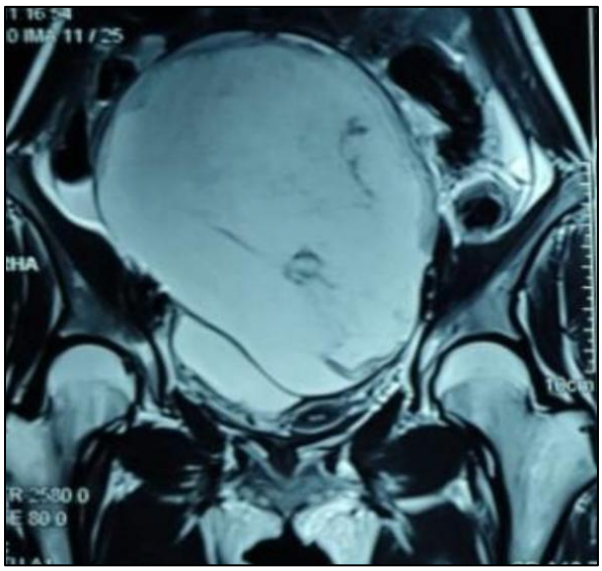

Figure 1: Large heterogenous multilocular, multiseptated, cystic mass arising from right ovary.

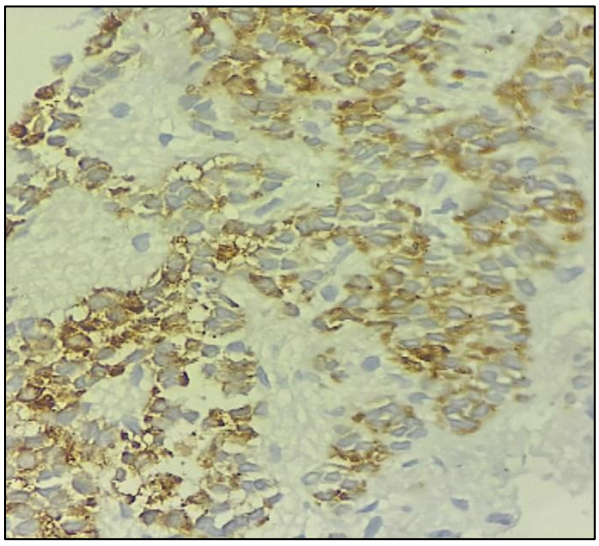

Figure 2: Juvenile granulosa cell tumor cells exhibited positive expression for inhibin on immunohistochemistry (X400 magnification).

The patient was scheduled for laparotomy with right salpingo-oophorectomy (RSO), which was intraoperatively converted to a fertility-sparing staging laparotomy with infracolic omentectomy and multiple peritoneal biopsies. Intraoperative findings included mild free fluid sent for cytology, a large trilobed adnexal mass of 15×10×18 cm with one lobe ruptured intraoperatively, and the mass was sent for a frozen section, which was suggestive of a borderline surface epithelial tumor.

Retroperitoneal spaces were opened bilaterally, revealing no enlargement of pelvic or para-aortic lymph nodes, while the uterus, left fallopian tube, ovary, and omentum appeared normal. The peritoneum and abdominal organs were palpated, a bowel run was performed, and the appendix was normal, with final intraoperative staging as FIGO stage IC1. The final histopathology report confirmed a diagnosis of juvenile granulosa cell tumor with micro metastasis in omentum, staged as pT3aN0, with immunohistochemistry (IHC) showing positivity for inhibin (Figure 2), PR, calretinin, and WT1, and negativity for P63, CK7, CK20, synaptophysin, chromogranin, and ER. The postoperative period was uneventful, and the patient tolerated the procedure well. The case was discussed in a multidisciplinary tumor board, where a decision was made to proceed with postoperative adjuvant paclitaxel and carboplatin chemotherapy. The patient completed four cycles of adjuvant chemotherapy and has remained in remission for one year, currently undergoing three-monthly surveillance.

DISCUSSION

Granulosa cell tumors (GCTs) are rare sex cord-stromal tumors, accounting for 1–5% of all ovarian neoplasms, and are classified into adult and juvenile subtypes, which differ in histology, clinical presentation, and disease progression, with the adult type comprising 95% of cases, typically affecting women over 40 years, whereas the juvenile type is rarer (<5%) and primarily occurs in prepubescent females. Both adult and juvenile GCTs commonly present with abdominal pain and increasing girth, with tumor rupture leading to acute abdominal pain in 6–10% of cases, while juvenile GCTs are often hormonally active, with estrogen secretion causing precocious puberty in premenarchal girls and irregular uterine bleeding in reproductive-age women, and rare androgen-secreting variants may induce virilization, although associations with Maffucci syndrome and Ollier disease have been suggested but remain unconfirmed.[2] Our patient presented with abdominal pain without any menstrual irregularity or signs of virilization.

Histologically, juvenile GCTs exhibit irregular follicles, luteinization, and a high mitotic rate, distinguishing them from the adult type, which features call-exner bodies and grooved nuclei with low mitotic activity, while immunohistochemistry positivity for inhibin is a key diagnostic marker. Our patient presented with similar histopathology findings with immunohistochemistry positive for inhibin. Imaging findings are nonspecific, with CT and ultrasound typically revealing a large multiloculated cystic mass with solid components and septations, whereas MRI provides better tissue contrast,

with T1-weighted hyperintensity indicating intratumoral haemorrhage (seen in up to 71% of cases) and a T2-weighted sponge-like appearance representing solid and cystic areas, with endometrial thickening possibly observed due to estrogenic stimulation, and in 10% of adult-type GCTs, an association with endometrial adenocarcinoma warrants consideration for preoperative endometrial biopsy, while hematogenous metastases to the lungs, liver, and brain are possible, though lymphatic spread is rare (~8%).[3] Our patient also presented with CEMRI findings of a large multiseptated right abdominopelvic mass.

GCTs follow the FIGO staging system for ovarian tumors, guiding treatment decisions and recurrence risk assessment, with 90% of juvenile GCTs diagnosed at stage I, carrying a 5-year survival rate of 90–100%, and unlike adult-type GCTs, juvenile tumors rarely recur post-resection, but when they do, the course is often aggressive with early relapse and poor prognosis, while advanced-stage disease correlates with higher recurrence rates (41% in stage III-IV) and increased mortality (30%), even in cases without macroscopic residual disease.

Surgical resection is the primary treatment, with fertility-sparing surgery preferred in early-stage disease, while chemotherapy is reserved for advanced or recurrent cases, with cisplatin-based regimens demonstrating efficacy, and recent studies suggesting the combination of taxanes (e.g., paclitaxel) with platinum-based agents, while radiotherapy has shown no significant survival benefit. Our patient presented in stage III and received postoperative adjuvant paclitaxel and carboplatin. For stage I tumors, surgery alone is often curative, while survival rates drop for stage II (55–75%) and for stage III-IV (22–50%), with poor prognostic indicators including nuclear atypia, high mitotic activity, extracapsular extension, tumor rupture, and residual disease post-surgery. While recurrence in juvenile GCTs is uncommon, when it occurs, it is often within the first year, and although early follow-up for three years was once deemed sufficient, reports of late recurrences beyond four years post-surgery suggest that longer surveillance may be necessary, while tumor markers such as inhibin are useful for monitoring recurrence.[4]

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