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Large Ovarian Juvenile Granulosa Cell Tumor in an Adolescent A Case Report and Literature Review

Benchaaboune Kenza, Belhaj Yassine, Tazi Zineb, Jayi Sofia, Fdili Alaoui Fatimazahrae, Chaara Hekmat, My Abdelilah Melhouf

Department of Gynecology and Obstetrics II, Hassan II University Hospital, Sidi Mohammed Ben Abdellah University, Fez, Morocco

Corresponding author: Benchaaboune Kenza, e-mail: kenza.benchaaboune@usmba.ac.ma

***Abstract**: Juvenile granulosa cell tumors are rare sex cord stromal ovarian neoplasms, representing approximately five percent of all granulosa cell tumors. They usually occur in premenarchal girls and young women and may present with endocrine manifestations or mass effect. We report the case of a sixteen year old adolescent with a history of rheumatic pancarditis and early menarche, admitted for progressive abdominal distension of one month duration. Imaging showed a large unilateral solid cystic adnexal mass with limited intra abdominal fluid and no secondary lesions. Tumor markers including CA 125, carcinoembryonic antigen, CA 19 9, beta human chorionic gonadotropin and lactate dehydrogenase were normal, whereas alpha fetoprotein was slightly elevated. After preoperative ureteral double J stenting, surgical staging was performed. Exploration found a 25 cm solid cystic tumor arising from the right ovary. The patient underwent right salpingo oophorectomy, omentectomy, peritoneal fluid aspiration and multiple biopsies. Intraoperative rupture led to FIGO stage IC1. Histology confirmed a juvenile granulosa cell tumor confined to the right ovary with negative peritoneal cytology. Close surveillance was chosen and the patient was disease free at six months. This case highlights the diagnostic challenge of a giant adnexal mass in adolescence and the importance of fertility sparing staging surgery, careful pathological confirmation and long term follow up.*

Keywords: juvenile granulosa cell tumor; ovarian tumor; sex cord stromal tumor; adolescent; fertility sparing surgery; FIGO stage IC1

Table 1: Summary of the clinical, radiological and surgical features of the present case

Variable	Finding
Age and history	16 years; pancarditis secondary to rheumatic fever; menarche at 8 years with regular cycles
Presentation	Progressive abdominal distension for 1 month, preserved general condition, no digestive or urinary symptoms
Imaging	Solid cystic adnexal mass: ultrasound 13 x 10 cm; CT approximately 17.6 x 15.2 x 26.3 cm with limited intraperitoneal fluid and no secondary lesions
Tumor markers	CA 125, CEA, CA 19 9, beta hCG and LDH normal; AFP slightly elevated
Surgery	Right salpingo oophorectomy, omentectomy, peritoneal fluid aspiration and multiple biopsies
Pathology and stage	Juvenile granulosa cell tumor confined to the right ovary; negative peritoneal cytology; FIGO IC1 due to intraoperative rupture69
Outcome	Close surveillance; no clinical recurrence at 6 months

1. Introduction

Granulosa cell tumors are the most frequent malignant sex cord stromal tumors of the ovary. They are classified into adult and juvenile subtypes according to clinicopathological features, and the juvenile subtype accounts for a minority of cases. Juvenile granulosa cell tumors are typically diagnosed in children, adolescents and young women. Although these tumors are malignant, many are confined to one ovary at diagnosis and have a favorable outcome after complete surgical treatment.

The clinical presentation is heterogeneous. Hormone secreting tumors may reveal themselves through isosexual

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precocious puberty, breast development, menstrual disorders or endometrial stimulation, whereas non secreting tumors may be discovered because of abdominal distension, pain or a palpable pelvic mass. Imaging usually identifies a unilateral adnexal mass with solid, cystic or mixed solid cystic features, but the preoperative diagnosis remains difficult because tumor markers may be non specific or normal.

Because most patients are young, the therapeutic strategy must balance oncologic safety with fertility preservation. Fertility sparing surgery with complete staging is generally preferred in early stage disease, whereas adjuvant chemotherapy is considered for advanced, recurrent or selected high risk cases. We report an exceptional case of a giant juvenile granulosa cell tumor in a sixteen year old adolescent, focusing on diagnostic difficulties, surgical management and follow up considerations.

2. Case Presentation

A sixteen year old adolescent with a history of pancarditis secondary to rheumatic fever was admitted for progressive abdominal distension evolving over one month. Menarche had occurred at the age of eight years, and the patient reported regular menstrual cycles. There were no digestive or urinary symptoms, and the general condition was preserved. Abdominal examination found a distended abdomen with diffuse tenderness on palpation. The main clinical features are summarized in Table 1.

Suprapubic pelvic ultrasonography showed a large mass measuring approximately 13 x 10 cm, with both solid and cystic components that were difficult to characterize. Contrast enhanced computed tomography performed for extension assessment demonstrated a very large unilateral adnexal solid cystic mass, measuring approximately 17.6 x 15.2 x 26.3 cm, with a heterogeneously enhancing solid component, limited intraperitoneal fluid and close relationships with vascular, digestive and urinary structures. No secondary localization was identified. Serum CA 125, carcinoembryonic antigen, CA 19 9, beta human chorionic gonadotropin and lactate dehydrogenase were within normal limits, while alpha fetoprotein was slightly elevated.

After placement of a double J ureteral stent, the patient underwent surgical staging. Intraoperative exploration revealed a 25 cm solid cystic tumor arising from the right ovary and extending toward the upper abdomen. Accidental intraoperative rupture of the cystic component occurred, with evacuation of abundant infected cystic fluid. The procedure consisted of right salpingo oophorectomy, omentectomy, aspiration of peritoneal fluid and multiple peritoneal biopsies. Because intraoperative tumor rupture was reported, the tumor was staged as FIGO IC1.

The final pathological examination concluded to a juvenile granulosa cell tumor limited to the right ovary. Peritoneal cytology was negative. After multidisciplinary discussion, close postoperative surveillance was retained. At six months after surgery, the patient was clinically in remission.

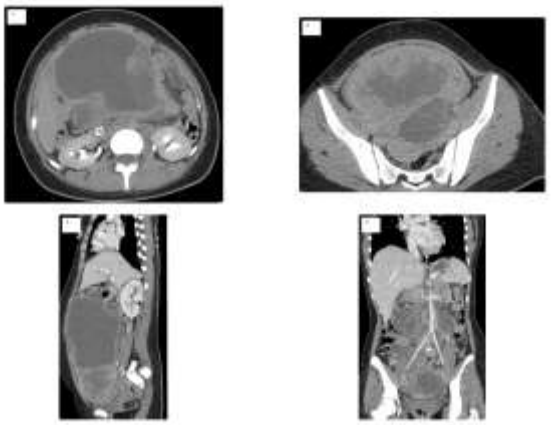

Fig. 1. Preoperative computed tomography showing a giant predominantly cystic solid cystic adnexal mass with a heterogeneously enhancing solid component on axial, sagittal and coronal views.

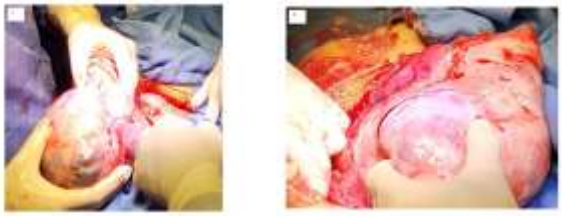

Fig. 2. Intraoperative appearance of the giant ovarian solid cystic tumor before surgical removal.

3. Discussion

Juvenile granulosa cell tumor is a rare ovarian neoplasm within the sex cord stromal tumor group. The juvenile subtype is generally observed in the first decades of life and is classically considered more aggressive than the adult subtype. However, it is frequently unilateral and confined to the ovary at diagnosis. This explains the favorable prognosis reported for most early stage cases after complete resection.

The present observation illustrates a common diagnostic challenge: the clinical picture was dominated by abdominal distension related to the size of the tumor, without digestive or urinary symptoms and without marked elevation of conventional epithelial or germ cell tumor markers. Normal CA 125 does not exclude juvenile granulosa cell tumor, and the role of CA 125 in this disease remains limited. Inhibin, anti Mullerian hormone and calretinin may be useful in diagnosis or follow up when available, because they are more closely related to granulosa cell differentiation.

Radiologically, juvenile granulosa cell tumors may appear as solid, cystic or mixed solid cystic adnexal masses. In our patient, the very large size and solid cystic architecture required computed tomography to evaluate extension and surgical relationships. The discordance that may occur between imaging suspicion and definitive origin highlights the importance of careful intraoperative assessment and final pathological confirmation.

Surgery remains the cornerstone of treatment. In adolescents and young women, fertility sparing surgery with unilateral salpingo oophorectomy and appropriate staging is generally accepted when the disease appears unilateral and resectable. Staging should include exploration of the peritoneal cavity, peritoneal washing or fluid cytology, omental assessment and biopsy of suspicious lesions. Systematic lymphadenectomy and routine biopsy of a normal contralateral ovary are not routinely recommended in early stage disease.

Our patient was classified as FIGO IC1 because of intraoperative rupture. The benefit of adjuvant chemotherapy in stage IC disease remains debated, especially when rupture is intraoperative, cytology is negative and complete resection has been achieved. Recent series suggest that stage I disease, including selected stage IC cases, may have a good outcome with surveillance alone, whereas advanced stage, residual disease and relapse are associated with poorer outcomes. The decision should therefore be individualized through multidisciplinary discussion, considering stage, completeness of surgery, histological features, patient age and follow up reliability.

Long term surveillance is essential because recurrence, although uncommon in early stages, can occur. Follow up usually combines clinical examination, pelvic imaging and tumor markers when initially elevated or when granulosa specific markers are available. In the present case, close surveillance was selected and the patient remained in remission at six months, but prolonged follow up is mandatory.

4. Conclusion

Juvenile granulosa cell tumor should be considered in any young patient presenting with a unilateral adnexal mass, even when conventional tumor markers are normal and imaging suggests benign or indeterminate features. This case is notable for the giant tumor size, the adolescent age of the patient and the FIGO IC1 classification following intraoperative rupture. Fertility sparing staging surgery remains central in management, while the indication for adjuvant treatment must be discussed individually. Careful pathological confirmation and prolonged follow up are required to detect recurrence early.

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Conflict of Interest

The authors declare no conflict of interest.

Consent for Publication

The manuscript uses anonymized clinical data. Written consent for publication should be confirmed by the authors before journal submission according to the journal policy.

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