

ADULT GRANULOSA CELL TUMOR OF THE OVARY: A CASE REPORT***Dr. Bouterfas M., Bennani A., Serroukh Mohamed M., Yousfi M. and Bargach S.**

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ABSTRACT

Adult-type granulosa cell tumors (AGCTs) belong to the sex cord-stromal group of ovarian tumors and account for 2-5% of ovarian neoplasms. Histopathology remains the gold standard for making a diagnosis of GCTs. Recent evidence has shown association of this tumour with Forkhead box L2 gene mutations. Surgery is the mainstay of treatment. Chemotherapy and/or radiotherapy are considered in patients with advanced stage or with unresectable recurrent disease. Granulosa cell tumors are well known for late recurrence and the stage at time of diagnosis is the only prognostic factor that is unequivocally related to survival.

INTRODUCTION

Granulosa cell tumours (GCT) are uncommon neoplasms, accounting for 2-5% of all ovarian carcinomas.^[1]

Rokitansky (1859) was the first to describe granulosa cell tumor of the ovary as a single tumor entity.^[2]

Although adult granulosa cell tumor (AGCT) can present at any age, it typically occurs around menopause, suggesting that the perimenopausal hormonal changes contribute to its pathogenesis.^[3]

Hyperoestrogenism reported in patients with GCT is related to tumor production of oestrogens, anti-Mullerian hormone (AMH), and inhibin B.^[4] According to histological findings, two different subtypes of GCT were identified: adult (AGCT) and juvenile (JGCT). AGCTs are more frequent.^[5]

CASE REPORT

A 52-year-old peri-menopausal woman (gravida 2, para 2), with no significant pathological history presented with a two years history of pelvic mass. Clinical examination found a pelvic mass lateralized to the left with 7cm long axis. Ultrasound (fig1) shows a left latero-uterine mass measuring 13 cm long axis, vascularized on Doppler suggesting an ovarian tumor. On MRI (fig2) heterogeneous right supra-latero-uterine mass suspicious of malignancy most probably of ovarian origin. The CA 125 was negative. The patient underwent a left adnexectomy by laparotomy, peritoneal cytology, right ovary biopsy and epiploic biopsy. The histopathological results showed: morphological and immunohistochemical profile consistent with AGCTs. Based on the histological findings a total hysterectomy with right adnexectomy and omentectomy were performed. The suites were simple, currently under clinical supervision.

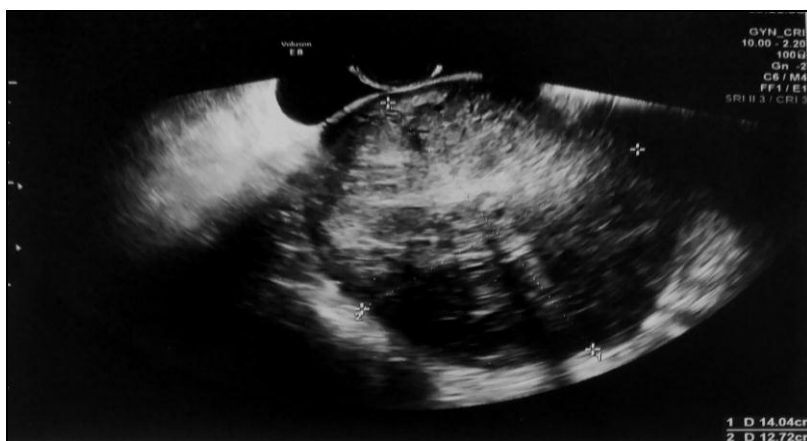


Figure 1: Ultrasound shows a left latero-uterine mass with heterogeneous echogenicity.

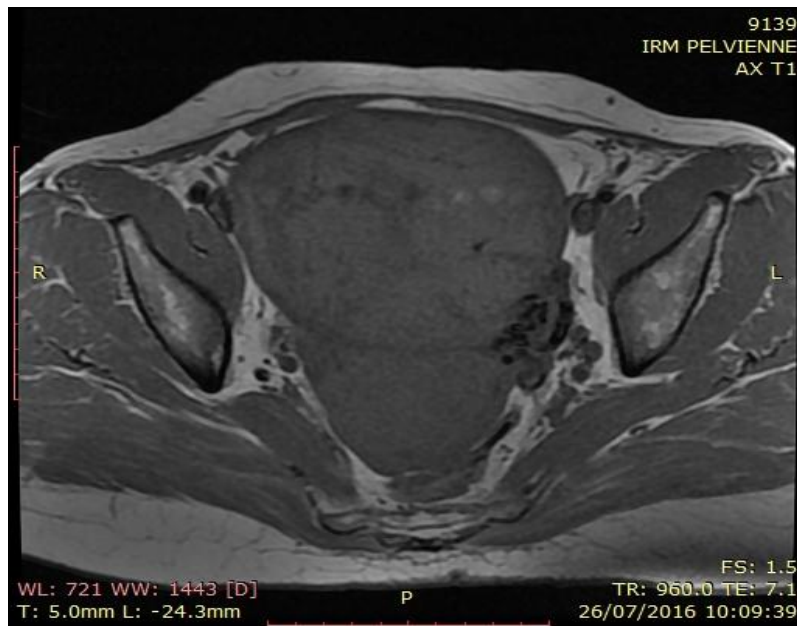


Figure 2: Images of MRI showing heterogeneous right supra-latero-uterine mass.

DISCUSSION

Granulosa cell tumor (GCT) is a malignant tumor originating from the sex-cord stromal cells of the ovary and represents 2% to 5% of all primary ovarian cancers.^[6] Approximately 5% occur before puberty (juvenile GCT) and the majority (95%) of the cases are the adult type of GCT (occur in people of reproductive age and postmenopausal age).^[7]

Adult granulosa cell tumors are usually considered low grade malignant tumors with a tendency to remain localized and demonstrate an indolent clinical course and late relapse.^[8]

The most reported signs in the literature are abdominal pain and/or abdominal distension (30% to 50%) and hormonal event such as postmenopausal bleeding, amenorrhea, and intermenstrual bleeding.^[9]

The differential diagnosis for GCTs should include other cystic masses and malignant tumors of the ovary (e.g., follicular cysts, cystadenoma, carcinoid, adenocarcinoma, and ovarian thecoma). In ultrasonography, the appearance of AGCT varies from cystic to solid, although the most common presentation is a solid and cystic mass with occasional hemorrhagic fluid.^[10]

MRI could provide useful information in accurately diagnosing OGCT (Ovarian granulosa cell tumor). The heterogeneous signal intensity on both T1WI (T1 weighted imaging) and T2WI (T2 weighted imaging) and high signal intensity on DWI images are more suggestive of OGCT diagnosis. ADC (Diffusion weighted imaging) value might be useful in differentiating OGCT from Other sex-cord stromal tumor.^[11]

On histological examination, the presence of Call-Exner bodies and “coffee-bean” nuclei with a low mitotic activity are typical characteristics of AGCTs.

Several studies have reported that approximately 70–97% of adult granulosa cell tumors carry a somatic c.402C>G missense point mutation in the FOXL2 gene.^[12] This makes FOXL2 a highly sensitive and relatively specific marker for adult granulosa cell tumors, as well as a useful diagnostic test for distinguishing between adult and juvenile granulosa cell tumors types and other sex cord- stromal tumors.^[13]

The mainstay of treatment is a complete surgery (hysterectomy with bilateral salpingo-oophorectomy) with staging for early stage and debulking surgery for advanced stage or recurrent disease.^[14]

Recurrence is predominantly located intraperitoneally.^[15] To date, an adjuvant chemotherapy for patients with completely (macroscopic) removed granulosa cell tumor of the ovary is not obligatory.^[16] Some authors recommend polychemotherapy containing cisplatin, vinblastine and bleomycin for patients with advanced stage or relapse.^[15]

and radiotherapy may also represent a treatment option for selected cases.^[15] The most important prognostic factor in this tumor types is the tumor stage.^[17]

The standard follow-up of AGCT patients typically includes regular gynecologic examinations with pelvic/transvaginal ultrasound and serum tumor markers. As in preoperative evaluation, the most sensitive and specific serum markers are inhibin B and AMH, which are also elevated in relapsed settings and correlate with disease burden.^[18]

CONCLUSION

Granulosa cell tumor is an uncommon ovary neoplasm. Disease stage seems to be the only reliable prognostic factor.

Granulosa cell tumors are well known for late recurrence. As a clinical consequence, life long follow-up for all patients with granulosa cell tumors is warranted and should not end after five years.

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