MEDICAL FORUM

A RARE CASE OF ADULT-TYPE GRANULOSA CELL TUMOR OF THE OVARY

Vu Dang Luu, Le Phuoc Dat**, Nguyen Thi Thu Hang***

SUMMARY

Adult granulosa cell tumor of the ovary (AGCT) is a rare malignant ovarian neoplasm, which represents 2% of all ovarian tumors and is the most common sex cord-stromal tumor of the ovary [1–3]. It is also associated with clinical estrogenic manifestations and is usually diagnosed between 50 and 54 years of age [4]. Although it shows malignancy, the AGCT usually demonstrates a better prognosis than the other types of ovarian cancer. In this article, we present a case report of an early AGCT with the features of ultrasound, CT scan, and MRI.

Keywords: *Adult granulosa cell tumor of the ovary, ovary, MR imaging, surgery.*

I. INTRODUCTION

Adult granulosa cell tumor of the ovary (AGCT) is a rare malignant ovarian neoplasm with an incidence of less than 3.7 per 100,000 persons, which represents 2% of all ovarian tumors and is the most common sex cord-stromal tumor of the ovary [1–3], [5]. It is usually divided into two subsets according to clinical and histological characteristics, namely, adult-type GCT (AGCT) and juvenile GCT (JCGT). The former is the main, which accounts for approximately 95% of all GCTs [6]. It can occur in women at any age but is usually detected between 50 to 54 years of age, mainly in the postmenopausal period [5], [7].

AGCT is often hormonally active, which leads to the prominent clinical features closely associated with clinical estrogenic manifestations, such as metrorrhagia, postmenopausal bleeding, and endometrial hyperplasia/ carcinoma.

Surgery is the primary treatment for ovarian AGCT. Meanwhile, other therapy (radiotherapy, chemotherapy, hormonotherapy, and targeted therapy), is applied for patients, who need adjuvant therapy, with advanced and recurrent disease [5], [6].

The prognosis of AGCT is favorable, especially the early stage, with the 5-year and 10-year survival rates for FIGO stage I being more than 90% and 84%, respectively [6], [7]. Meanwhile, that of patients with stage III or IV disease was only 22-50%. Fortunately, most patients are diagnosed with AGCT at an early stage (78-91%), with a mean diameter of approximately 11cm (range 0.5-30cm) [5], [7]. However, the importance of early diagnosis of AGCT is undeniable, and radiology is one of the most crucial tools for screening AGCT. Therefore, in this article, we will present an early AGCT case study and focus on radiographic findings on ultrasound and MRI.

II. CASE REPORT

A 57-year-old woman was admitted to the hospital because of 2-week pelvic pain, without digestive disorder or abnormal vaginal bleeding. She was married for 17 years. PARA 2012. She was in a postmenopausal period

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for 7 years. She did not use any hormonal contraceptives or ovulation-inducing drugs. There was no family history of a similar illness. She was non-diabetic, non-hypertensive, non-smoker, and non-alcoholic.

On general examination, rest all vitals being within normal limits, the patient complained of pelvic pain without tenderness point. Speculum examination revealed cervicitis and on bimanual examination, the clinician found a painless mass, which has a smooth surface with good mobility in the right adnexa.

Complete blood count, renal and liver function tests, urine, and peripheral smear examination were all normal. Laboratory examination results were within normal limits with tumor marker beta-HCG value < 0.1 U/mL; AFP: 1.4 ng/mL; C125: 13 U/mL; CEA: 2.54, HE4: 34.2.

On transabdominal ultrasound, there is a heterogenous isoechoic mass at the pelvis which is 34x39mm in size, with a regular border, isoechoic to the adjacent uterus, and mildly increased vascularity. According to the above feature, it can be categorized as ORADS US 4 lesion.

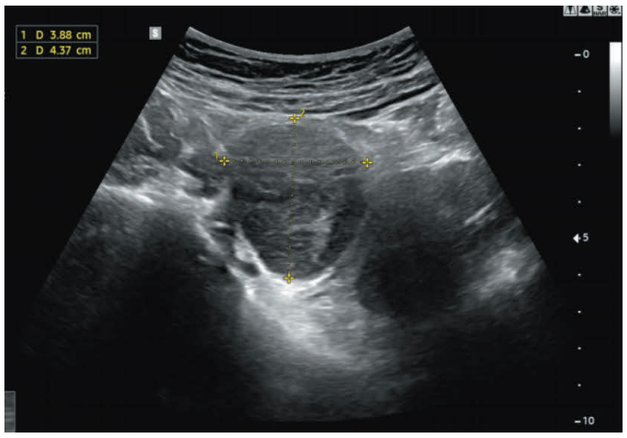

Figure 1. A homogeneous isoechoic mass (*) in the right adnexal area, 44x39mm in size, with a regular border.

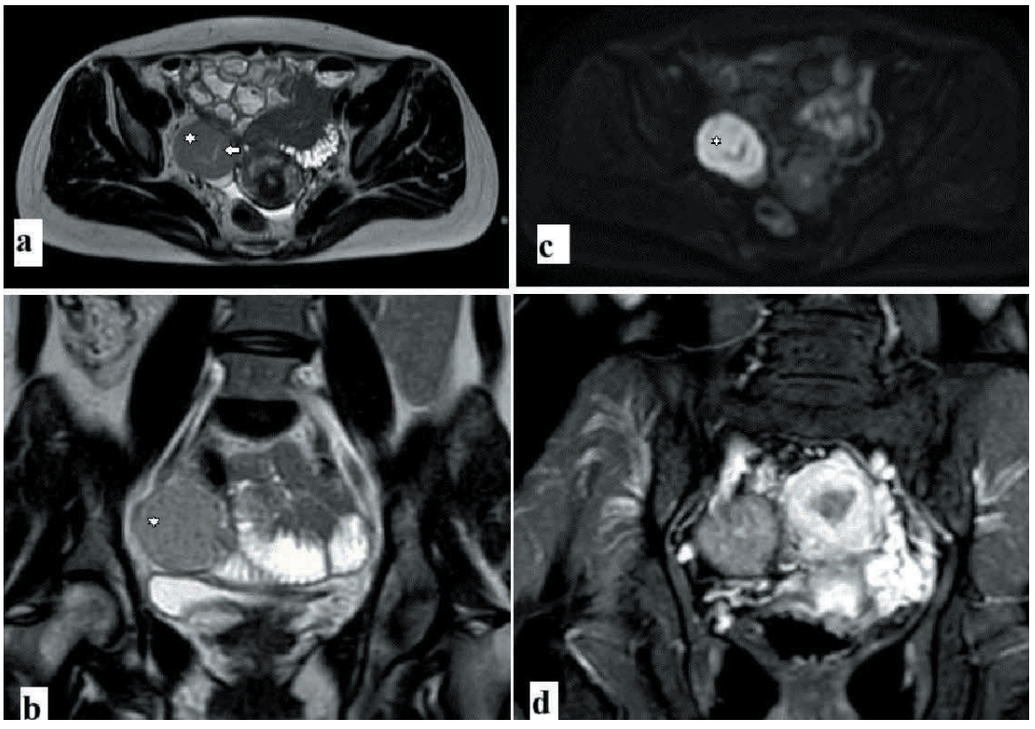

Figure 2. *(a, b) A homogeneous intermediate ovarian mass (*) on T2W images, with a regular border. There aren't any signals suggesting cystic or hemorrhagic components. The vascular origin from the right internal iliac artery goes into the tumor (arrow), which showed restricted diffusion on DWI (c) and ADC (not shown). (d)After contrast, the tumor is mildly enhanced, which is lower enhanced compared to the uterus.*

The patient underwent a pelvic MRI which confirmed a right ovarian solid mass (Figure 2). Some diagnosis is considered including AGCT, fibroma, thecoma, Brenner tumor, or dysgerminoma.

Besides, the endometrial uterus thickness is increasing for a postmenopausal woman, approximately 9.7mmmm, suggesting a high estrogenic level, which may be associated with the tumor, favor the diagnosis of AGCT or thecoma.

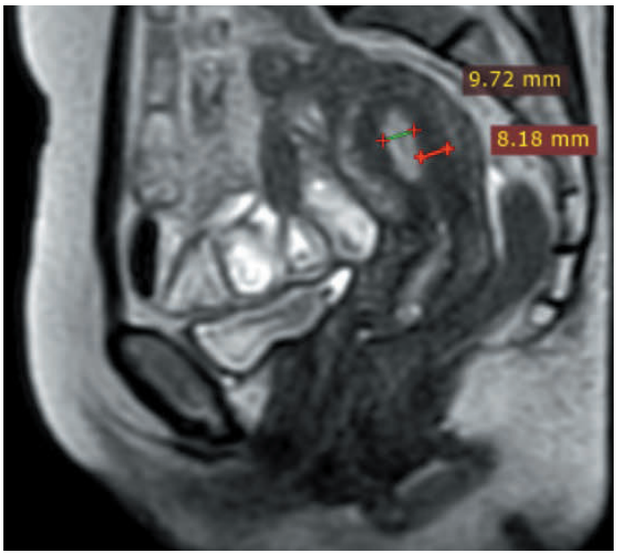

Figure 3. The thickening of endometrium is approximately 9.7mm (normally lower than 5mm for postmenopausal women).

Additionally, there is no infiltration around the mass or abnormal lymph nodes are seen. For staging the tumor more carefully, the whole-body CT scan, and breast ultrasound are indicated, which revealed normal results. Therefore, the IA stage is appropriate for this situation.

Then, a total abdominal hysterectomy, bilateral salpingo-oophorectomy, and omentectomy were performed and confirmed the right adnexal solid mass, which did not invade adjacent organs on macroscopy.

Macroscopic, the tumor was solid, soft, and yellowish, and no papillary growths or cystic structures were found. Some small patchy adenomyosis is also found, and there are no remarkable findings upon gross examination of the fallopian tube or left ovary.

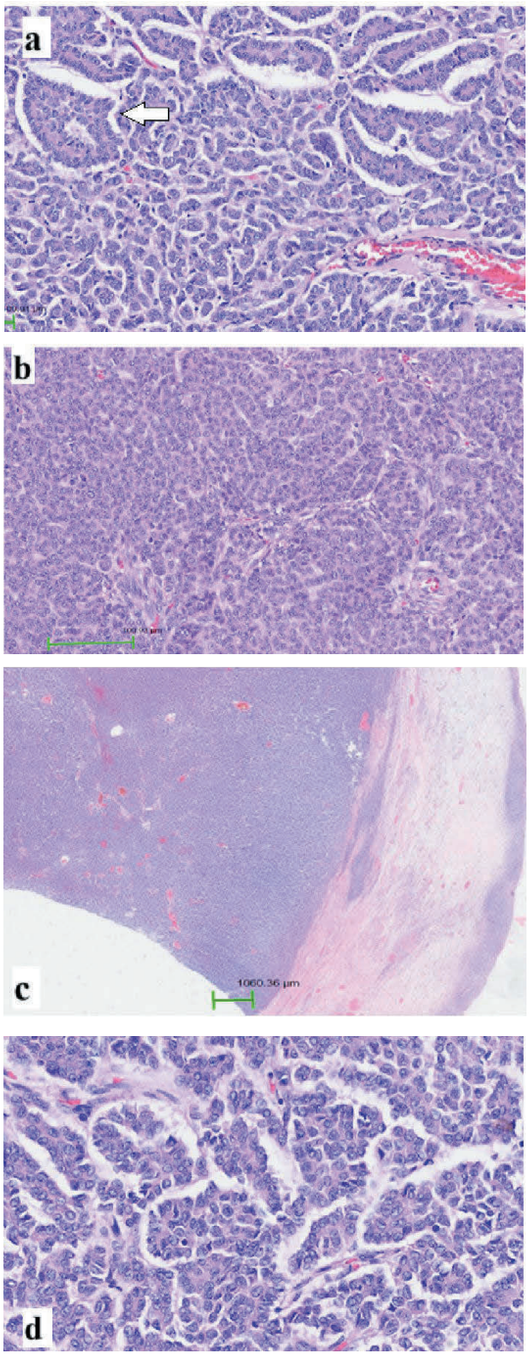

Figure 4. *(a), (b), (d): Relatively uniform tumor cells with pale, grooved nuclei (coffee has been nuclei). (b): Diffusing, trabecular growth pattern of the tumor cell. (a) Cell-Exner bodies with small follicle-like structures filled with eosinophilic material. (c) The boundary of the tumor is integrity, without invading the remaining normal ovary.*

Microscopic, the tumor tissue was arranged in a solid, diffuse, and trabecular pattern of the tumor cells with scant cytoplasm and pale, uniform angulated, and grooved nuclei. Some structures suggest microfollicular form (resembling Call-Exner bodies with small follicle-like structures filled with eosinophilic material). The Thin-wall capillaries and stroma cells also are found in the tumors. The boundary of the tumor is integrity, without invasive tumor cells into the remaining normal right ovary, which supports preoperative IA staging. The diagnosis of AGCT is established. Additionally, the hyperplasia of the uterus myometrium and adenomyosis is confirmed.

III. DISCUSSION

Granulosa cell tumors are one type of ovarian sex-cord tumor and account for one of the largest groups of hormonally active ovarian tumors, which are classified into adult and juvenile types. In contrast to the former, which usually affects women older than 30 years, the latter occurs mainly in children [1], [3–5].

Clinically, the most common symptoms of AGCT in postmenopausal patients are abnormal vaginal bleeding and a unilateral ovarian mass. Meanwhile, irregular vaginal bleeding, amenorrhea, is the main expression in premenopausal women, and rarely infertility [5–7]. Abdominal pain may be related to the compression or the torsion of the tumor. However, approximately 20% of the patients are asymptomatic at the time of initial diagnosis [5].

There is a spectrum of AGCT radiographic findings, which is classified into five patterns including multilocular cystic, thick-walled single locular cystic, thin-walled single locular cystic, homogeneous solid, and heterogeneous solid types, according to Ko et al. [1]. Subsequently, Kim et al. simplified the classification based on the two most commonly observed types as multi-septal cystic lesions and non-lobulated solid lesions of internal cystic lesions [3], [5].

Histopathologically, AGCT has variable histologic patterns, correlated to radiologic morphology, because of the different arrangement of tumor cells, including microfollicular, macrofollicular, trabecular, insula, water-silk, solid-tubular, diffuse, and luteinized patterns [1]. It is characterized by uniform pale, longitudinal grooves nuclei tumor cells (coffee bean nuclei). The Call-Exner bodies can be found, as well [1]. The morphologic appearances are also affected by intra-tumoral hemorrhage or cystic degeneration [1], [5].

The homogeneous solid pattern in this case is uncommon, and appeared in 2/13 and 1/11 patients, in Ko et al. and Kim et al. reports, respectively [1], [3]. On histopathology examination, these solid tumors were composed of evenly distributed trabeculae of granulosa cells interspersed by fibrous stromal components, disorganized sheets of granulosa cells without any cystic change, or hemorrhage [1]. It has been reported that this solid morphology is usually small, around 5cm, but it was relatively large in Ko et al. report (7cm and 9cm) [1], [3], [7]. These above features are matching with our case study, which is appropriate for the diagnosis of an AGCT homogeneous solid pattern.

Additionally, previous studies not only revealed uterine enlargement and endometrial thickening in some cases of AGCT (30-80% [1], [3], but also the uterine changes unusual for the patient's age, such as adenomyosis, prominent and thickened junctional zone, and increased signal intensity of myometrium [3].

IV. CONCLUSION

AGCTs have variable morphology depending on different histopathology. The most common appearances are multi-septal cystic lesions and non-lobulated solid lesions of internal cystic lesions. Homogeneous solid is an unusual pattern of AGCT, without any intra-tumoral hemorrhage or cystic degeneration. Estrogenic manifestations, with abnormal vaginal bleeding or endometrial thickening, may suggest a hormonal active tumor, for which AGCT mainly accounts.

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