Ovarian granulosa cell tumor – case report and literature review

Amelia Milulescu1,2, Cristian Gabriel Viisoreanu2, Nicolae Bacalbasă3, Irina Balescu4, Tiberiu-Augustin Georgescu5,6, Roxana Elena Bohiltea3,7, Claudia Stoica8,9

1Department of Obstetrics and Gynecology, Faculty of Medicine, “Titu Maiorescu” University, Bucharest, Romania
2Breast Institute, Monza Oncology Hospital, Bucharest, Romania
3Department of Obstetrics and Gynecology, “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania
4Department of Visceral Surgery, Ponderas Academic Hospital, Bucharest, Romania
5Department of Pathology, “Carol Davila” University of Medicine and Pharmacy Bucharest, Romania
6“Alessandrescu-Rusescu” National Institute for Mother and Child Health, Bucharest, Romania
7Department of Obstetrics and Gynecology, Filantropia Clinical Hospital, Bucharest, Romania
8Department of Anatomy, “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania
9Department of Surgery, Ilfov County Emergency Hospital, Bucharest, Romania

ABSTRACT

Gigantic (>10 cm) ovarian cysts are a rare finding in gynecology. In literature, there are only a few cases of giant ovarian cysts reported, mainly in older patients. Granulosa cell tumor (GCT) of the ovary is frequently a hormonally active, stromal cell neoplasm with the ability to produce sex steroids, such as estrogens. We report the case of a 61-year-old postmenopausal woman with a giant left ovarian cyst measuring 39 x 28 cm. The excision of the giant left ovarian cyst was performed, with intraoperative histologic examination; the result was granulosa cell tumor. We continued the surgery with a total abdominal hysterectomy and bilateral salpingo-oophorectomy and complete staging. On the final histopathological report the result was granulosa cell tumor of the ovary. The management of these high-risk ovarian tumors should have a strict follow up in order to avoid complications.

Keywords: ovarian cancer, granulosa cell tumor, ovary

INTRODUCTION

Ovarian sex cord stromal tumors (SCST) are a group of benign and malignant neoplasms that develop from the sex cord, stromal cells or both. Some ovarian SCSTs produce steroid hormones, androgens or estrogens, and therefore may present with signs of estrogen excess or virilization [1].

Ovarian sex cord tumors are more infrequent than tumors of epithelial cell and germ cell origin. Malignant ovarian sex cord tumors account for less than 8 percent of ovarian neoplasms. They most commonly have a low grade with rare lymph node metastases, and a good prognosis [2].

Clinical characteristics for the adult granulosa cell tumor are most commonly found in middle-aged and older women (median age 50 to 54 years) and comprise 95% of these neoplasms. The appearance of the ovarian mass is variable; usually large (mean diameter 12 cm but may be as large as 30 cm), tan or yellow. The ultrasound reveals, usually unilateral, echogenic, septated cystic, or solid mass. May look like a mucinous cystadenoma or be filled with serous fluid or clotted blood, and can associate ascites. Signs of excess of estrogens are found in over 50% of patients, and endometrial hyperplasia/uterine bleedings in 25 to 50%. It can sometimes also associate endometrial cancer [3,4].

Corresponding author:
Roxana Bohiltea
E-mail: r.bohiltea@yahoo.com

Article History:
Received: 4 December 2021
Accepted: 30 December 2021
CASE PRESENTATION

A 61-year-old postmenopausal patient presented to our department with complaints of massive abdominal distention which started gradually 3 months ago. The patient also complained of vaginal bleeding and difficulty in breathing, with no other gastrointestinal or urinary symptoms. No family history of malignant ovarian and/or breast cancer in first-degree relatives have been found.

The patient's abdomen was heavily distended, tense on palpation. Blood analysis revealed hemoglobin of 10.8 g/dl, and 332,000/μl platelets. CA-125 was slightly elevated (48.9 U/ml) and ROMA score was 33.05% (high risk of ovarian cancer). Because of the associated uterine bleeding, the patient underwent a curretage for endometrial biopsy, and the result was endometrial hyperplasia, with no atypical findings.

An abdominal and pelvis CT scan was also performed. The CT revealed a huge pelvi-abdominal mass measuring 29 x 15 x 22.4 cm, which originated from the left ovary (Figure 1), mesenteric and pelvic lymph node enlargement, bilateral ureterohydronephrosis and pelvic ascites.

Because of elevation of ROMA score and CA-125 levels, the result of the CT scan, ovarian malignancy was suspected. The patient underwent exploratory laparotomy with removal of the left ovarian mass with intraoperative hystologic examination (Figure 2) – with the result of ovarian carcinoma, most probably granulosa cell tumor.

The surgery was completed with total hysterectomy and bilateral salpingo-oophorectomy and complete staging.

There were no intra or postoperative complications. On day 4 of surgery, patient was discharged. Her final histopathology report was adult granulosa
cell tumor limited to the left ovary with negative lymph nodes, ascites with no tumoral cells.

**DISCUSSION**

Rokitansky described granulosa cell tumors for the first time in 1855 [5]; he described them in concordance with their appearance, adjacent to the granulosa cells of ovarian follicles. The incidence of sex cord stromal tumors is 0.2/100,000 patients [6], with an average age at diagnosis of 50 years. The symptoms are can be specific or not and range from: abdominal pain to abdominal distension related to mass effect and clinical manifestations related to the hormonal profile due to the estrogenic and androgenic effects (intermenstrual bleeding, postmenopausal bleeding or amenorrhea), this justifying why the GCTs are frequently associated with endometrial hyperplasia or endometrial adenocarcinoma [7], thus endometrial biopsy is essential.

There are many factors that can determine the prognosis, the most important prognostic variable being the stage of the disease. Almost 75% of patients with adult granulosa cell tumor of the ovary are at stage I, with the lesion being limited to the ovary [8].

The recommended treatment consists in complete surgery (hysterectomy with bilateral salpingo-oophorectomy), with complete staging or debulking surgery in case of advanced or recurrent disease [9].

GCTs have a tendency for late recurrence. The recurrence rate in a study made by Dridi et al. on 31 patients was 32%, most of them being located in the pelvis [10].

**CONCLUSIONS**

GCT of the ovary is a very rare neoplasm, with a very good prognosis. Due to its known potential of relapsing after many years, it is very important for the patient to have a rigorous follow-up.

**Conflict of interest:** none declared

**Financial support:** none declared

**REFERENCES**

1. Busquets M, Gonzalez-Bosquet E, Muchart J, Rovira C, Lailla JM. Granulosa cell tumor and endometrial cancer: a case report and review of the literature. *Eur J Gynaecol Oncol*. 2010;31(5):575-8.
2. Thrail MM, Paley P, Pizer E, Garcia R, Goff BA. Patterns of spread and recurrence of sex cord-stromal tumors of the ovary. *Gynecol Oncol*. 2011 Aug;122(2):242-5.
3. Al-Hussaini M, Al-Othman Y, Hijazi E, McCluggage WG. A Report of Ovarian Sertoli-Leydig Cell Tumors With Heterologous Intestinal-type Glands and Alpha Fetoprotein Elevation and Review of the Literature. *Int J Gynecol Pathol*. 2018;37:275.
4. Lim D, Oliva E. Ovarian sex cord-stromal tumours: an update in recent molecular advances. *Pathology*. 2018;50:178.
5. Björkholm E, Silfverswärd C. Prognostic factors in granulosa-cell tumors. *Gynecol Oncol*. 1981;11:261-274.
6. Quirk JT, Natarajan N. Ovarian cancer incidence in the United States, 1992-1999. *Gynecol Oncol*. 2005 May;97(2):519-23.
7. Bompas E, Freyer G, Vitrey D, Trillet-Lenoir V. Granulosa cell tumour: review of the literature. *Bull Cancer*. 2000 Oct;87(10):709-14.
8. Zhao D, Zhang Y, Ou, Z, et al. Characteristics and treatment results of recurrence in adult-type granulosa cell tumor of ovary. *J Ovarian Res*. 2020;13:19.
9. Kottarathil VD, Antony MA, Nair IR, Pavithran K. Recent advances in granulosa cell tumor ovary: a review. *Indian J Surg Oncol*. 2013 Mar;4(1):37-47.
10. Dridi M, Chraiet N, Batti R, et al. Granulosa Cell Tumor of the Ovary: A Retrospective Study of 31 Cases and a Review of the Literature. *Int J Surg Oncol*. 2018;2018:4547892.