CASE REPORT

Surgical Staging and Recurrence Management of Ovarian Granulosa Cell Tumor

B. Díaz-de la Noval, M. Choqueneira Dionicio
Gynaecology and Obstetrics Department, University Hospital La Paz, Madrid-Spain

Article Information

Received: 13 January 2016
Accepted: 22 March 2016
Plagiarism software: Turnitin

Keywords:
Granulosa cell tumor,
Recurrence,
Surgical staging,
Surgical management

Díaz-de la Noval

ABSTRACT

Introduction: Ovarian Granulosa Cell Tumors (GCT) is a potentially malignant tumor. Accurate diagnosis requires histological and immunohistochemical study. The treatment of choice is surgical staging and debulking staging according to International Federation of Gynecology and Obstetrics (FIGO) criteria, preferred by laparoscopy. The therapeutic role of adjuvant therapy is unclear. Relapses arefrequent, usually late and insidious. **Case Report:** we report two cases of Ovarian Granulosa Cell Tumor. First case, a laparoscopic oophorectomy for complex adnexal cyst, intraoperative study does not differentiate between Granulosa Cell Tumor or clear cell carcinoma of the ovary, so we performed a complete staging surgery by laparoscopy. Second case aparaortic recurrence twenty years after first surgery. Non-steroidal anti-androgen therapy was useful to control progression of the disease, finally laparoscopic excision of the lesion. **Conclusion:** A complete surgery in the management of Granulosa Cell Tumor is recommended. Relapses can occur very late and response to chemotherapy is poor, so it is important to perform a careful initial staging or debulkingprocedure because intraoperative pathological diagnosis is difficult and confusing.

INTRODUCTION

Ovarian Granulosa Cell Tumors (GCT) is a potentially malignant tumor. Its diagnosis is made by histology after surgical excision and staging according to International Federation of Gynecology and Obstetrics (FIGO) criteria, being the most important prognostic factor. It is a tumor with good prognosis, 90% are diagnosed at stage I; metastases rarely occur. The 5-year survival for stage I patients is above 90%, 55-75% in Stage II and 20-50% in Stage III. Although, large tumor size and breakage are associated with worse prognosis and risk of recurrence. Relapses may be delayed beyond 10 to 20 years of diagnosis.[1] This brief review reports two cases from the Gynecology Oncology unit at University Hospital La Paz, Madrid.

Access this article online	
Website: www.actamedicainternational.com	**Quick Response code**
DOI: 10.5530/ami.2016.2.37	

CASE REPORT

Our first and most recent case is a 78-year old woman with an abdominal mass and wasting syndrome. She was hysterectomized at age 42 for fibroids. Abdominal tomography diagnosed a 15cm pelvic tumor mass depending on left ovary and a 5cm mucinous cystic lesion in the pancreas tail (biopsy showed haemorrhagic content) and bilateral hydronephrosis, no ascites, lymphadenopathies or carcinomatosis. Tumor markers were above the limit. Suspecting malignancy we performed a decisional laparoscopy. We observed a large encapsulated tumor mass, retroperitoneal, growing up from the left ovary. No lymphadenopathy was observed. Intraoperative study of the mass was a GCT or clear-cell tumor (Figure 1). Complete staging surgery was followed by hysterectomy, right adnexectomy, omentectomy, peritoneal biopsies, pelvic and para-aortic lymphadenectomy by laparoscopy. The pathology study confirmed GCT, being a FIGO stage

Corresponding Author:
Begoña Diaz de la Noval, Gynaecology and Obstetrics Department, University Hospital La Paz, Madrid-Spain. Tel: +34616306566,
E-mail: begodelanoval@gmail.com

IA. No adjuvant treatment was indicated, currently free of disease.

Second case is a 59-year old woman diagnosed of a GCT tumor and underwent an incomplete surgery (only ovarian cystectomy by laparotomy) in a different centre in 1995, FIGO stage IC, without adjuvant therapy, multiple surgeries for abdominal relapse. In 2008 she first came to our institution, she had a multiple peritoneal relapse treated with a complete debulking surgery by laparotomy and adjuvant chemotherapy (6 courses of carboplatin-paclitaxel). In July 2014 appears again a 25mm retroperitoneal relapse so she started chemotherapy with Orteronel till May 2015. Tomography shown persistence of a 45mm retroperitoneal mass next to the aortic bifurcation, the patient referred muscle weakness in the lower limbs associated with compression of lumbar roots L4-L5 (Figure 1). We performed a laparoscopic extraperitoneal paraortic lymphadenectomy with removal of the cystic lesion located at left infrarenal level. 32 tumor-free lymph nodes were obtained, cystic lesion containing multiple tumor nests and extracapsular infiltration that supports GCT variety adult. No adjuvant therapy, currently free of disease.

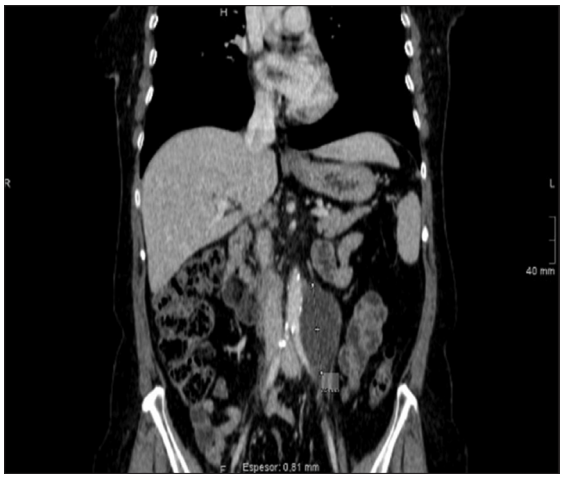

Figure 1: Tomography image showing a 16x45 mm left Para-aortic mass

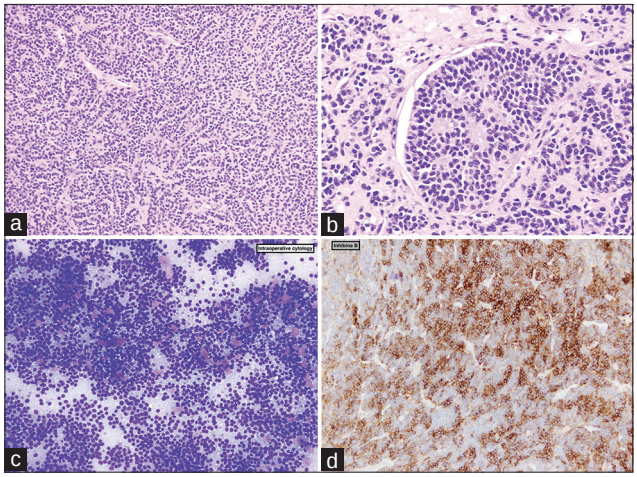

Figure 2: (a) Histological appearance of a granulosa cell tumor. Atypical small pale round cells in an alveolar conf guration. (b) Histological appearance of a granulosa cell tumor. Few mitosis per high-power f eld. Sheets of cells with small nuclei and central slot feature shaped coffee beans (Call-Exner bodies). (c) Histological appearance of a granulosa cell tumor. Intraoperative cytology. Atypical small pale round cells in an alveolar conf guration. (d) Histological appearance of a granulosa cell tumor. (Haematoxylin-eosin and Inhibina-A positive stains)

DISCUSSION

GCT is a sex cord-stromal ovarian tumor, more frequent in peri-menopausal age group. Most cases of GCT are detected in early stages, as discussed. The conventional treatment remains complete surgical excision. In young women with stage IA fertility preservation is possible after discarding concomitant endometrial cancer or hyperplasia due to the hyperestrogenism hormonal stimulation caused by the tumor.[2] Also due to the rarity of lymph node metastases in the initial diagnosis, it is suggested that the pelvic and paraortic lymphadenectomy can be omitted as part of the surgical staging.[3,4] The malignant potential of these tumors is in the high frequency of relapses, which can occur very late, and poor response to chemotherapy.

The gross appearance is that of a unilateral, hard, yellow and highly vascular mass. The characteristic growth pattern of well differentiated Granulosa Cells (less than ten mitoses per high-power field), small, uniform, pale, round or oval, with small nuclei and central slot feature shaped coffee beans. These tumor cells are surrounded by abundant stromal component forming small cystic areas of fluid and cellular debris called Call-Exner bodies, found in 50% of GCTs. Inhibina-A is a high sensitive and specific biomarker of GCT and prognostic factor in the ovary (Figure 2a-d).[5]

Because it is difficult to distinguish GCTs from epithelial ovarian tumors pre-operatively, specially clear cell carcinoma, and there is a low-accuracy of intraoperative pathological diagnosis, the surgical procedure would be the same as suspected of epithelial ovarian cancer.[1]

Regarding adjuvant therapy, there is no standard treatment. Some centres recommend platinum based regimens,[2,3] others suggest therapy only when residual disease after surgery or in recurrences, as with the second patient presented in this report.[3] Recent reviews suggest that BEP (bleomycin plus etoposide and cisplatin), GnRH analogues or antiestrogen therapy is an effective regimen after surgery in stages over IC, this is also recommended for recurrent disease after surgical excision.[1] Nevertheless, surgical resection was essential in handling both cases, actually without adjuvant therapy and free of disease.[6]

The malignant potential of GCT is in the high frequency of relapses, which can occur very late, and poor response to chemotherapy. A complete surgery in the management of Granulosa Cell Tumor is recommended. Beyond surgical excision as primary treatment there is no standard way for managing recurrences in Granulosa Cell Tumor. It is important to perform a careful initial staging or debulking procedure because intraoperative pathological diagnosis is difficult and confusing. However, the knowledge of the potential long-term relapses in these tumors makes it necessary to establish treatment protocols and clinical studies with large case series.

REFERENCES

1. Ryousuke T, et al. Presentation of two patients with malignant granulosa cell tumors, with a review of the literature. World J Surg Oncol, 2012;10(1): 185-9.
2. Park JY, Long Jin K, Kim DY, Kim JH, Kim YM, Kim KR, Kim YT, Nam JH. Surgical staging and adjuvant chemotherapy in the management of patients with adult granulosa cell tumors of the ovary. Gynecol, 2012; 125(1): 80-6.
3. Meisel JL, et al. The role of systemic chemotherapy in the management of granulosa cell tumors. Gynecol Oncol, 2015; 136(3): 505-11.
4. Sekkate S, et al. Ovarian granulosa cell tumors: a retrospective study of 27 cases and a review of the literature. World J Surg Oncol, 2013; 11(1):142-7.
5. González-Díaz E, Fernández Fernández C, Santos Salas Valién J, González García C. Adult granulosa cell tumor of the ovary. Prog Obstet Gynecol, 2008;51(10):593-604.
6. Ud Din N, Kayani N. Recurrence of adult granulosa cell tumor of the ovary: experience at a terciary care center. Ann Diag Pathol, 2014;18:125-8.

How to cite this article: Noval BD, Dionicio MC. Surgical staging and recurrence management of ovarian granulosa cell tumor. Acta Medica International.. 2016;3(2):185-187.

Source of Support: Nil, **Conflict of Interest:** None declared.