

Retroperitoneal granulosa cell tumor

Marie Claire Vassallo, Mandy Collict, Tiffany Buhagiar,
Mark Formosa

ABSTRACT

A rare case of a retroperitoneal granulosa cell tumor (GCT) is presented in a 52-years-old menopausal woman who was referred with an incidental finding of an asymptomatic pelvic mass. Extraovarian granulosa cell tumors are derived from ectopic gonadal tissue which is situated along the embryonal route of the genital ridge. An ultrasound revealed a heterogenic mass with increased vascularity on colour doppler.

Keywords: Extraovarian, Granulosa cell tumor, Retroperitoneal

How to cite this article

Vassallo MC, Collict M, Buhagiar T, Formosa M. Retroperitoneal granulosa cell tumor. J Case Rep Images Obstet Gynecol 2019;5:100045Z08MV2019.

Article ID: 100045Z08MV2019

doi: 10.5348/100045Z08MV2019CR

INTRODUCTION

Granulosa cell tumors (GCTs) are uncommon, sex cord-stromal tumors which account for 2-5% of ovarian neoplasms [1]. GCTs are composed of both granulosa, theca cells and fibroblasts in varying quantities or can consist of almost entirely granulosa cells [2]. GCTs are known to recur years after the initial diagnosis or metastasize after initial treatment [3]. Primary GCTs occurring at extraovarian sites are rare, however it is even rarer to have the primary tumor arising from the retroperitoneum [4].

Retroperitoneal tumors occur with an incidence of 2–6% with an 80–85% chance of malignancy [2]. Retroperitoneal tumors most often originate from the pancreas, kidneys and adrenals [4].

CASE REPORT

A 52-year-old menopausal woman who was referred for a palpable right adnexal mass detected on routine vaginal examination. A transvaginal scan revealed a right adnexal complex mass of mixed echogenicity measuring 6.6×5.7×8 cm (Figures 1 and 2) with increased vascularity on colour doppler (Figure 3). The patient had no medical or surgical history of note.

CA125 was 5.8 U/ml (reference range up to 30.2 U/ml), lactate dehydrogenase 220 U/l (reference range up to 220 U/l). Surgical removal of the tumor was offered to the patient. A decision for laparotomy was made due to the relatively large and solid nature of the mass.

At operation, the uterus was atrophic and both ovaries and tubes were unremarkable. The right ovary was suspended by a small fold of peritoneum to a 12×8 cm retroperitoneal mass. A routine total abdominal hysterectomy and bilateral oophorectomy was performed. A vascular surgeon was asked to assist with mobilisation of the retroperitoneal lesion as this was exactly over the internal iliac vessels. The lesion revealed a white, fragmented, soft interior which was sent for histological examination. The lesion was completely excised, its pedicle was ligated and the underlying iliac vessels were examined for patency. The patient made an uneventful recovery.

Marie Claire Vassallo¹, Mandy Collict¹, Tiffany Buhagiar², Mark Formosa¹

Affiliations: ¹Department of Obstetrics & Gynaecology, Mater Dei Hospital, Malta, Dun Karm Street, Msida, Malta; ²Department of Pathology, Mater Dei Hospital, Malta.

Corresponding Author: Dr. Mandy Collict (Higher Specialist Trainee), Department of Obstetrics & Gynaecology, Mater Dei Hospital, Malta; Email: Mandy.caruana@gov.mt

Received: 09 March 2019

Accepted: 25 March 2019

Published: 22 April 2019

Histologically, tubes and ovaries were unremarkable. However, sections from the pelvic mass revealed tumor composed of cuboidal to polygonal cells arranged predominantly in call-exner bodies, trabecular, solid and insular patterns. Occasionally, the cells had a coffee bean nucleus with central grooves. The tumor cells strongly and diffusely expressed inhibin, vimentin and calretinin (Figure 4). Broad range cytokeratin revealed patchy positivity. CK7, CK20, ER, EMA, CD99, synaptophysin and chromogranin A were not expressed. The proliferative

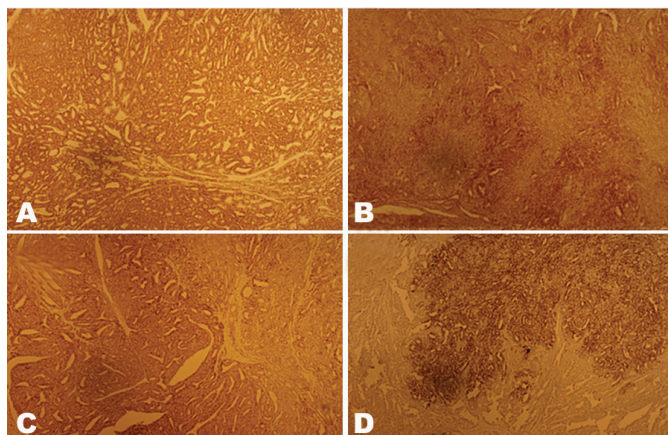


Figure 4(A–D): The immunohistochemical staining results. Immunohistochemical expression was positive for (A) Vimentin (B) Calretinin (C) Inhibin and (D) BRCK.



Figure 1: Right adnexal mass of mixed echogenicity on transvaginal scan.

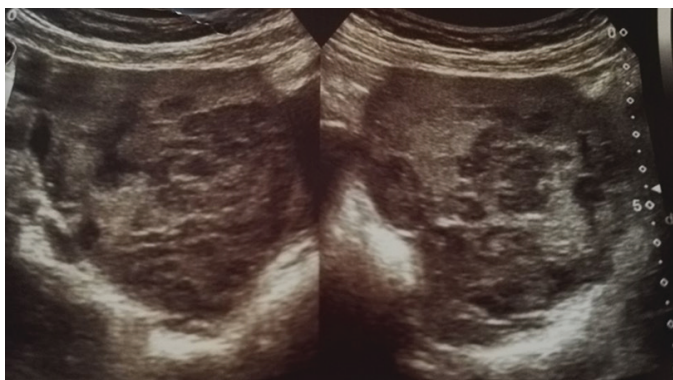


Figure 2: Heterogenic mass consisting of cystic and solid components on transvaginal scan.



Figure 3: The lesion with increased vascularity on ultrasound scan.

index (Ki-67) was low (5%). From the above mentioned findings, a diagnosis of an extra ovarian granulosa cell tumor was made.

The patient made an uneventful recovery and she was discharged home three days after her procedure. The case was discussed at the departmental multidisciplinary team meeting and was referred to the oncology unit where she is being followed up by regular tumor markers including CA125, CEA, CA19-9, FSH, LH, LDH and Inhibin. Ultrasounds of the pelvis at three months and nine months and then every four months in her second year and serial imaging with computed tomography and magnetic resonance imaging.

DISCUSSION

Granulosa cell tumors (GCTs) constitute 70% of ovarian sex cord-stromal tumors, capable of secreting sex steroids like oestrogen [5]. GCTs can occur at any age and can be of the adult (95%) or juvenile (5%) type, subtypes distinguished by clinical and histological features [4]. Commonly these tumors present during the perimenopause or early menopause, the median age of diagnosis being 50-54 years of age [5].

The less common juvenile subtype, occurs in the first three decades of life [1] and manifests with either isosexual precocious pseudopuberty or pelvic/ abdominal pain due to a large pelvic mass. Rarely, androgen-secreting GCTs give rise to virilising features. GCT is a vascular tumor which may rupture resulting in an acute abdomen; giving rise to abdominal pain, haemoperitoneum and hypotension, mimicking an ectopic pregnancy in younger individuals [5].

Tumor derived oestrogen may give rise to abnormal vaginal bleeding patterns – menorrhagia, metrorrhagia and postmenopausal bleeding (up to 50%) [6]. The latter results from prolonged exposure of the endometrium to oestradiol, giving rise to endometrial hyperplasia or

endometrial adenocarcinoma. GCTs can present with endometrial adenocarcinoma, the latter present in 5-10 % of cases [7]. On the other hand, endometrial carcinoma can be an incidental finding in a case of suspected GCT [5]. In adults, GCT has been associated with cystic disease of the breast [8].

It is necessary to follow up all patients with a history of GCTs both clinically and biochemically with tumor markers, because 17% of relapses occur more than 10 years after the initial diagnosis was made. The pelvis is a common site for recurrence, patients' post-oophorectomy can develop GCT from an extraovarian location. Metastasis can occur years after initial treatment. It is important to rule out metastasis (excluding previous GCT of the ovary) before a diagnosis of an extraovarian GCT is made [3].

Extraovarian GCTs are very rare indeed, and have been documented to develop in the broad ligament, retroperitoneum, mesentery, omentum, liver and adrenals [3]. It has been proposed that the histogenetic origin of extraovarian GCT derives from ectopic gonadal stromal tissue originating from the mesonephros [3, 9]. A dual origin from both the mesonephros and coelomic epithelium has also been suggested [10]. The mesonephros seems to be fundamental for the development of the sex-cord; this might suggest the sites of the extraovarian GCTs being limited to the retroperitoneum, the adrenals or the broad ligament; all of which differentiate in close proximity to the mesonephros and the mesonephric duct [10].

CONCLUSION

Granulosa cell tumors must always be followed-up closely due to the possible risk of recurrence. Granulosa cell tumors need to be considered in the differential diagnosis of retroperitoneal tumors of the pelvis. The dissection of any retroperitoneal tumor will require the assistance of a surgeon with necessary skills to completely risk of perform it as there is increased malignancy.

REFERENCES

1. Chou CF, Huang WC. Granulosa cell tumor of the ovary. *Ci Ji Yi Xue Za Zhi* 2016;28(2):84-5.
2. Chiriac D, Todorut FM, Fogarassy A, Corpade A, Grigoras D. P31.04: Primary retroperitoneal granulosa cell tumor: A case report. *Ultrasound in Obstetrics & Gynecology* 2014;44(Suppl 1):181-365.
3. Maniwadekar MR, Patil NJ. Extraovarian granulosa cell tumor of mesentery: A case report. *Patholog Res Int* 2010;2010:292606.
4. Vasu PP, Leelamma JP, Mohammed BA, Yesodharan J. Primary granulosa cell tumor of retroperitoneal origin: A rare presentation with emphasis on cytomorphology. *J Cytol* 2016;33(1):52-4.
5. Schumer ST, Cannistra SA. Granulosa cell tumor of the ovary. *J Clin Oncol* 2013;21(6):1180-89.

6. van der Vorst MJ, Bleeker MC, Boven E. Unusual presentation of granulosa cell tumor of the ovary. *J Clin Oncol* 2010;28(29):e554-6.
7. Evans AT 3rd, Gaffey TA, Malkasian GD Jr, Annerggers JF. Clinicopathologic review of 118 granulosa and 82 theca cell tumors. *Obstet Gynecol* 1980;55(2):231-8.
8. Kim SH, Park HJ, Linton JA, et al. Extraovarian granulosa cell tumor. *Yonsei Med J* 2001;42(3):360-3.
9. Paul PC, Chakraborty J, Chakrabarti S, Chattopadhyay B. Extraovarian granulosa cell tumor. *Indian J Pathol Microbiol* 2009;52(2):231-3.
10. Motta PM, Makabe S. Germ cells in the ovarian surface during fetal development in humans. A three-dimensional microanatomical study by scanning and transmission electron microscopy. *J Submicrosc Cytol* 1986;18(2):271-90.

Author Contributions

Marie Claire Vassallo – Conception of the work, Design of the work, Acquisition of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mandy Collict – Conception of the work, Design of the work, Acquisition of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Tiffany Buhagiar – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mark Formosa – Conception of the work, Acquisition of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

Copyright

© 2019 Marie Claire Vassallo et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

Access full text article on
other devices



Access PDF of article on
other devices





INTERNATIONAL JOURNAL OF
CASE REPORTS AND IMAGES



VIDEO JOURNAL OF
CLINICAL RESEARCH



VIDEO JOURNAL OF
BIOMEDICAL SCIENCE



INTERNATIONAL JOURNAL OF
HEPATOBIILIARY AND
PANCREATIC DISEASES



INTERNATIONAL JOURNAL OF
BLOOD TRANSFUSION AND
IMMUNOHEMATOLOGY



EDORIUM JOURNAL OF
OPHTHALMOLOGY



Submit your manuscripts at
www.edoriumjournals.com



EDORIUM JOURNAL OF
MEDICINE



EDORIUM JOURNAL OF
CARDIOTHORACIC AND
VASCULAR SURGERY



JOURNAL OF CASE REPORTS
AND IMAGES IN ORTHOPEDICS
AND RHEUMATOLOGY



EDORIUM JOURNAL OF
PSYCHOLOGY



EDORIUM JOURNAL OF
CELL BIOLOGY



JOURNAL OF CASE REPORTS AND IMAGES IN
DENTISTRY



EDORIUM JOURNAL OF
CANCER



EDORIUM JOURNAL OF
PSYCHIATRY



JOURNAL OF CASE REPORTS AND
IMAGES IN INFECTIOUS DISEASES



EDORIUM JOURNAL OF
ANATOMY AND EMBRYOLOGY



EDORIUM JOURNAL OF
SURGERY



JOURNAL OF CASE REPORTS
AND IMAGES IN PATHOLOGY



EDORIUM JOURNAL OF
ANESTHESIA