Retiform Sertoli-Leydig Cell Ovarian Tumour Presenting with Psammoma Bodies: A Rare Case Report

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ABSTRACT

Sertoli-Leydig cell tumours (SLCT) are rare sex cord-stromal neoplasms accounting for less than 0.2% of all ovarian tumours. We report a case of a patient who was operated due to a solid-cystic mass in the lower abdomen and the pathology report confirmed retiform variant of SLCT. A 24-year virgin female patient, with no preexisting health anomaly, drug use, or surgical history, presented with abdominal distension that started about 2 months ago. According to the FIGO 2018 classification, the final pathology report showed a stage IIIC SLCT and after oocyte cryopreservation, adjuvant chemotherapy was planned for this patient. The management of SLCT with retiform pattern is difficult because of the rarity of the tumour. Further prospective studies are needed to optimise the management of SLCT.

Key Words: *Retiform type, Sertoli-Leydig cell tumours, Sex cord-stromal tumour, Ovary.*

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INTRODUCTION

Sertoli-Leydig cell tumours (SLCT) are rare sex cord-stromal neoplasms accounting for less than 0.2% of all ovarian tumours.[1] The mean age at occurrence for SLCT is 25 years, whereas the average age at diagnosis for retiform SLCT is 15 years. SLCT typically consists of androgen-producing cells, so there are signs of virilisation due to androgen production in about half of the cases. However, virilisation is less common in retiform pattern SLCT. Although SLCT generally have good outcomes, grade and stage are of great importance in the prognosis. Approximately 100% survival has been reported in well-differentiated tumours. However, poorly differentiated tumours, tumours with heterologous mesenchymal elements, and rupture of the tumour have been associated with adverse outcomes.[2] Excessive amount of estrogenic activity (menometrorrhagia, postmenopausal bleeding) is rarely seen in patients.[3]

We, herein, report a case of a young female patient who was operated due to a solid-cystic mass in the lower abdomen and whose pathology report confirmed retiform variant of SLCT.

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CASE REPORT

A 24-year virgin female patient, with no preexisting health anomaly, drug use, or surgical history, presented with abdominal distension for about 2 months. Physical examination showed no signs of virilisation or hirsutism. A cystic structure of 12×10 cm with solid components was detected by ultrasonography in the lower abdomen and pelvis. Pelvic magnetic resonance imaging (MRI) was also planned for this patient. On MRI, a large abdominopelvic cystic mass with solid components seemed to arise from lower abdomen, measuring 14.7×11.2 cm. The mass showed irregular heterogeneous enhancement and local diffusion restriction in postcontrast series. Minimal fluid was detected in the pelvis. CA 125 elevation (94.7 IU/ml *vs*. normal, 35 IU/ml) was detected whereas other tumour markers (CEA, 1.26 ng/ml; CA 19-9, 7.28 IU/ml; CA 15-3, 15.28 IU/ml; beta HCG, 0.1 mIU/ml; AFP, 1.33 ng/ml) were within normal levels.

The patient was informed about the surgery and a written informed consent was obtained. Exploratory laparatomy was performed with a midline incision below the umbilicus. Right ovary, right tube, and uterus were found to be normal during exploration.

About 100 cc of free fluid was detected in the abdomen and it was aspirated for cytological examination. A solid-cystic mass, measuring approximately 15 cm and originating from the left ovary, was found. Left salpingoophorectomy was performed without rupturing the mass and material was sent for frozen section consultation. Malignant-benign distinction could not be confirmed by frozen section.

Thereupon, omentectomy, appendectomy, and bilateral pelvic lymph node sampling were performed. No other palpable pathological lesion was detected during abdominal exploration. However, biopsy samples were obtained from lesions that looked like possible intestinal metastatic implants. All metastatic implants were removed and no residual tumour was left behind.

Macroscopic examination of the left adnexal mass revealed a tumour measuring 17×14×9.5 cm. The outer surface was smooth. On sectioning, the tumour was yellow, had soft consistency, and included papillary-like structures. Histopathological examination revealed a tumour with wide finger-like projections and in some areas papillary structures lined by pseudostratified columnar cells. Finger-like projections and papillary structures were accompanied by various sized glands and slit--like tubules. In some areas of the tumour, retiform spaces and irregular glands with cuboidal cells were detected. In some areas, small calcifications with Psammoma bodies were seen. Immunohistochemical analysis showed that all tumour cells were positive for estrogen and progesterone receptors. Tumour cells also tested positive for WT1, CD56 and focal weak positive for pancytokeratin, EMA, vimentin and AFP. Tumour cells were negative for inhibin, *OCT3/4*, calretinin, CK7, CK20, PLAP, S100, synaptophysin and chromogranin. *P53* immunohistochemical staining was wild-type (Figure 1). Despite being tested negative for inhibin, diagnosis of retiform variant of SLCT was made based on the histopathological features and other immunohistochemical stains.

Left fallopian tube, uterus, appendix, pelvic lymph nodes and omentum were tumour-free. But, peritoneal implant was detected which was positive for malignant tumour, measuring 4.5×3×1.5 cm.

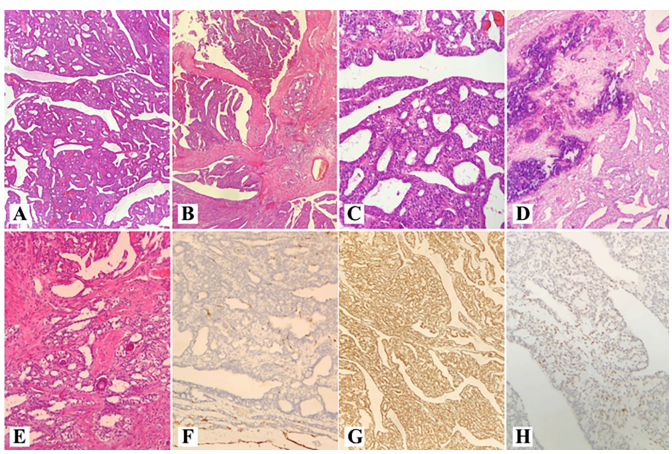

Figure 1: (A) Broad finger-like projections along with glands of various sizes and slit-like tubules (H&E, ×40). (B) Cystic spaces containing papillary structures on the left side, slit-like spaces within the fibrotic background on the right centre (H&E, ×40). (C) Tubular structures composed of pseudostratified columnar cells (H&E, ×200). (D) Wide calcification areas on the left side, slit-like tubular structures on the right side (H&E, ×40). (E) Slit-like spaces and concentric calcifications consistent with psammoma bodies (H&E, ×200). (F) Pancytokeratin is positive on the surface of epithelium, focally positive in tumour (×100). (G) Estrogen receptor expression in tumour cells (×40). (H) Wild-type staining of *p53* in tumour cells (×200).

According to the FIGO 2018 classification, the final pathology report showed the tumour to be a Stage IIIC SLCT and after oocyte cryopreservation, adjuvant chemotherapy was planned for this patient. The patient received 3 cycles of BEP therapy (bleomycin, etoposide, and cisplatin).

DISCUSSION

SLCT are generally rare and account for less than 0.5% of all primary ovarian neoplasms.[2] Retiform SLCT usually occur at a young age (mean age, 15 years) and present as large abdominal tumours and pain rather than virilisation. Recently, Sarkar *et al*. published 2 case reports about retiform variant of SLCT and it is noteworthy that both patients were reported to be 2 years old.[4] Yang *et al*. published a retrospective study about SLCT containing 80 cases, among which 59 (73.8%) patients were premenopausal and 21 (26.3%) were postmenopausal.[5] In an analysis of 15 patients with SLCT published by Wang *et al*., the average age was between 25 and 69 years and retiform variant of SLCT was detected in 5 out of 15 patients.[6]

Since SLCT are sex cord-stromal neoplasms, they are expected to test positive for inhibin. The data showed that some SLCT could be negative for inhibin as in the present case.[7] Leydig cells were found to be present in 44–98% of SLCT and they are strongly positive for inhibin.[8] In this case, Leydig cells were not identified. This might be related to the negativity for inhibin. Other markers were compatible to the previously published literature.[7,8] Besides, there was a case report presenting 2 cases of retiform variants of SLCT with psammoma bodies in the literature.[9] As far as we know, there is no such report in English literature after that. Therefore, to our knowledge, this is the third case of retiform variant of SLCT with psammoma bodies.

SLCT are uncommon sex cord-stromal tumours associated with both germline and somatic *DICER1* mutations, the frequency of which has varied widely in different studies (0 to 62.5%).[10] As a limitation of this study, *DICER1* could not be tested in the hospital. However, genetic screening and a test for germline *DICER1* mutation were recommended to the patient.

CONCLUSION

Management of ovarian SLCT is difficult because of their rarity. Retiform variant is a special subtype of SLCT. Further, prospective studies are needed to evaluate the course of cases with malignant forms of this tumour.

PATIENT'S CONSENT:

Informed, written consent was taken from the patient.

COMPETING INTEREST:

The authors declared no competing interests.

AUTHORS' CONTRIBUTION:

CYO: Conception and design, drafting and supervision.
MC: Analysis and interpretation of data.
NYC: Acquisition of data.
DTA: Critical revision of the manuscript.
GSY: Administration and technical support.

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