Case report

A GIANT OVARIAN SEX CORD STROMAL TUMOR PRESENTING AS A MASSIVE ABDOMINO-PELVIC MASS: A RARE CASE REPORT

ABSTRACT

Objective: To present a case report of ovarian spindle cell neoplastic lesion with sex cord stromal differentiation, highlighting diagnostic challenges in the absence of immunohistochemistry (IHC) .

Methods: Clinical, radiological, surgical, and histopathological data were reviewed for patient presenting with large abdomino-pelvic mass suspicious for ovarian spindle cell neoplasm. Diagnostic criteria and limitations due to lack of IHC were discussed.

Results: We report a rare case of a giant ovarian spindle cell neoplasm with sex cord-like differentiation in a 52-year-old hypertensive postmenopausal female presenting with a massive abdomino-pelvic mass. Imaging suggested a large complex adnexal tumor (O-RADS V) with elevated CA-125 levels. Staging laparotomy revealed a 32×24×21 cm right ovarian mass weighing over 15 kg with no evidence of metastasis. Histopathology revealed a spindle cell neoplasm with features suggestive of a sex cord–stromal origin.

Conclusion: Ovarian spindle cell neoplasms with sex cord stromal differentiation pose significant diagnostic challenges, especially when IHC is not feasible. Awareness of clinical, radiological, and histomorphological features along with literature guidance supports appropriate surgical management. Further research and improved access to IHC are essential for precise diagnosis and tailored therapy.

Keywords: Ovarian Spindle Cell, Neoplasms, Sex Cord, Stromal, Diagnostic Challenges

INTRODUCTION

Ovarian spindle cell neoplasms are rare tumors characterized histologically by spindle-shaped cells and often show sex cord stromal differentiation. These tumors encompass a heterogeneous group including pure stromal tumors like microcystic stromal tumor (MST) and sclerosing stromal tumor (SST), as well as mixed sex cord stromal tumors. Clinically, they may present as large abdomino-pelvic masses with nonspecific symptoms and elevated tumor markers such as CA-125 [1]. Imaging often shows complex solid-cystic masses with varying degrees of ascites, and they may be classified as high risk (O-RADS V) for malignancy. Histopathological diagnosis can be challenging due to overlapping features with other ovarian neoplasms, emphasizing the role of immunohistochemistry (IHC). However, in many settings, IHC may not be available, complicating definitive diagnosis and management decisions.

Sex cord–stromal tumors (SCSTs) of the ovary represent a heterogeneous group of neoplasms arising from the sex cords (granulosa, Sertoli, and Leydig cells) or ovarian stroma (fibroblasts and theca cells). They account for about 5–8% of all ovarian tumors and exhibit diverse histological patterns and clinical behaviours [1]. SCSTs include granulosa cell tumors, Sertoli-Leydig cell tumors, sclerosing stromal tumors, and microcystic stromal tumors among others [2].

These tumors often present with nonspecific symptoms such as abdominal distension, pelvic pain, or mass effect. Hormonal manifestations like virilization or estrogen excess are less frequent in older women [3]. Giant ovarian SCSTs (>20 cm) are exceedingly rare, with only a few cases documented in literature [4].

Histopathological evaluation is the cornerstone for diagnosis; however, due to overlapping morphology, immunohistochemistry (IHC) is often required to confirm lineage differentiation [5]. Common IHC markers include inhibin, calretinin, SF-1, WT-1, and β-catenin [6].

We report a rare case of a massive ovarian spindle cell neoplasm with possible sex cord-like differentiation, presenting as an abdomino-pelvic mass in a postmenopausal woman.

MATERIAL AND METHODS

This was a single case-based observational study conducted in the Department of General Surgery and Department of Oncology at a tertiary care teaching hospital conducted on a female patient presenting with large abdomino-pelvic mass suspicious for ovarian spindle cell tumor. Clinical data including age, comorbidities, symptoms, and tumor markers were collected. Imaging studies with ultrasound and contrast-enhanced CT were evaluated for tumor characteristics, size, ascites, and metastasis. Surgical data focused on staging laparotomy findings including tumor size, laterality, ascitic fluid volume, and presence of metastatic implants or lymphadenopathy. Histopathological examination characterized tumor morphology, and when available, differential diagnosis based on histomorphology was recorded. IHC was not feasible due to logistic constraints, diagnosis was based on morphology. Literature from the past ten years was reviewed to contextualize findings.

Case Presentation

A 52-year-old postmenopausal female, known hypertensive on regular medication, presented with progressive abdominal distension for six months. There was no history of weight loss, altered bowel habits, or menstrual abnormalities.

Clinical examination:

Abdomen was distended with a palpable, firm, non-tender mass measuring approximately 30×25 cm, extending from pelvis to epigastrium. P/V examination revealed fullness in the right fornix.

Investigations:

1. Ultrasonography (USG): Huge abdomino-pelvic mass (25×25 cm), complex solid-cystic lesion; O-RADS V.
2. Contrast-enhanced CT (CECT) Abdomen: Mild ascites with a 30×25×20 cm complex right adnexal solid-cystic mass; no retroperitoneal lymphadenopathy or metastases.
3. CA-125: 234 U/mL (elevated).

Surgical findings:

A staging laparotomy was performed. Intraoperatively, about 500 ml of ascitic fluid was noted. A right adnexal solid-cystic mass measuring 32×24×21 cm and weighing over 15 kg was seen arising from the right ovary. No omental, peritoneal, serosal, hepatic, or splenic deposits were observed. Uterus, left adnexa, and retroperitoneal nodes were unremarkable.

Histopathology:

Sections revealed a spindle cell neoplastic lesion arranged in intersecting fascicles and whorls. On the basis of morphology, the following differential diagnoses were suggested:

1. Pure stromal tumor of ovary with sex cord-like differentiation (microcystic stromal tumor/sclerosing stromal tumor).
2. Mixed sex cord–stromal tumor.

Uterus, cervix, left adnexa, omentum, and peritoneal biopsies were negative for malignancy. Immunohistochemistry could not be performed due to logistical constraints.

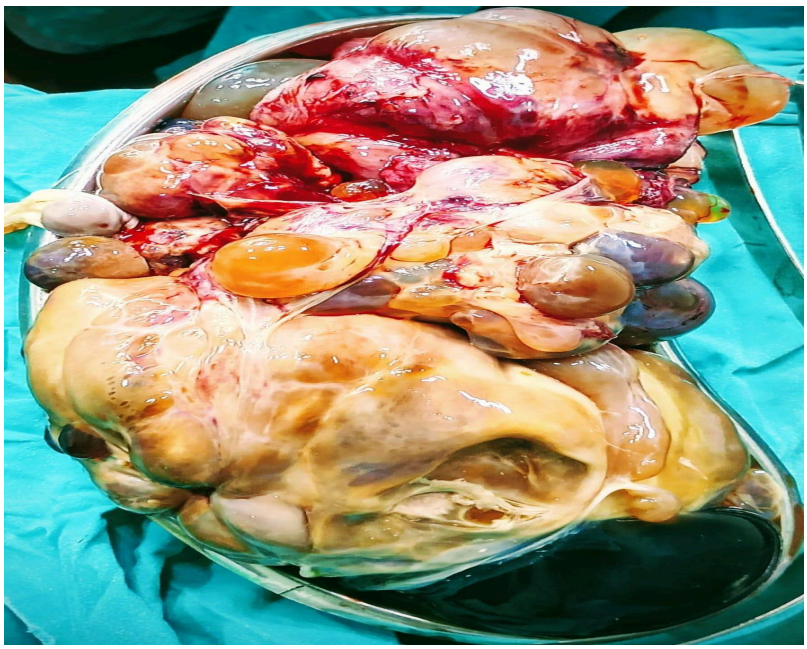

Figure 1: Giant ovarian stromal tumor, right adnexal solid-cystic mass of size 32x24x21 cm weighing more than 15 kg

DISCUSSION

Ovarian spindle cell neoplasms are uncommon and diagnostically challenging. The differential diagnoses in large solid-cystic adnexal masses with spindle cell morphology include pure stromal tumors such as microcystic stromal tumor (MST) and sclerosing stromal tumor (SST), as well as mixed sex cord stromal tumors. Microcystic stromal tumors, characterized by microcystic architecture and immunoprofile positive for CD10, vimentin, WT1, and β-catenin nuclear staining, are usually unilateral and present as large pelvic masses in young women. These tumors are low grade with good prognosis after surgical resection. Sclerosing stromal tumors, typically

affecting younger females under 30 years, demonstrate a benign clinical course with characteristic radiological features such as well-encapsulated, hypervascular solid-cystic masses showing avid contrast enhancement on MRI [2,3]. Surgical excision conserving ovarian function is preferred. Mixed sex cord stromal tumors represent a heterogenous group with variable clinical behavior and greater risk of recurrence in non-granulosa histologies and advanced disease stages. Retroperitoneal lymphadenectomy may be omitted as lymph node metastasis is rare.

Sex cord–stromal tumors are rare ovarian neoplasms with varied histologic subtypes and clinical manifestations. The age distribution is broad, ranging from adolescence to old age, though certain subtypes like granulosa cell tumors predominate in the peri- or postmenopausal age group [7].

The present case exhibited a large spindle cell tumor suggestive of stromal differentiation. Similar to our findings, Young et al. reported that stromal tumors can attain enormous sizes before diagnosis, often mimicking epithelial malignancies [8].

Microcystic stromal tumors (MSTs) are an uncommon subtype first described by Irving and Young in 2009, typically showing spindle to oval cells, microcystic areas, and β-catenin positivity on IHC [9]. Sclerosing stromal tumors (SSTs), on the other hand, usually occur in younger women and show a pseudolobular pattern with vascular prominence [10]. Distinction from fibromas, thecomas, and low-grade sarcomas may require immunoprofiling.

Elevated CA-125, though often associated with epithelial ovarian malignancy, can be mildly raised in large benign tumors due to peritoneal irritation or mechanical factors [11].

In this case, absence of metastasis and organ involvement, combined with localized spindle morphology, points toward a low-grade tumor. However, the lack of IHC limited definitive subtyping.

Similar cases of giant ovarian stromal tumors (>30 cm) have been rarely reported. Kusaka et al. described a 14 kg sclerosing stromal tumor in a 54-year-old woman [12], while Chen et al. (2023) reported a 12 kg microcystic stromal tumor successfully managed by complete excision [13]. These reports emphasize that despite massive size, SCSTs usually have favorable prognosis after surgical removal.

Prognosis:

Most sex cord–stromal tumors are stage I at presentation and have excellent outcomes following complete excision [14]. Adjuvant therapy is rarely required unless malignant features are confirmed. Regular follow-up is essential for early detection of recurrence.

The role of IHC in refining diagnosis of ovarian spindle cell and sex cord stromal tumors is critical, helping distinguish subtypes based on markers such as α-inhibin, calretinin, WT1, and others. However, limitations exist including logistic issues and occasional overlapping IHC profiles, so morphologic evaluation remains important [15,16].

This case report highlights the challenge when IHC is unavailable, where diagnosis relies heavily on histomorphology and clinical correlation. Large size, solid-cystic complex nature, ascites without metastasis, and spindle cell morphology should raise suspicion of sex cord stromal tumors. Careful surgical staging, tumor resection, and multidisciplinary discussion are essential for optimal management. Further research and improved access to IHC are needed to improve diagnosis and guide prognosis and therapy in ovarian spindle cell neoplasms.

This case underscores the diagnostic challenge of large ovarian spindle cell tumors and highlights the importance of considering stromal neoplasms in the differential diagnosis of giant ovarian masses.

CONCLUSION

This case illustrates a rare presentation of a giant ovarian spindle cell neoplasm with sex cord-like differentiation. Even though such tumors can mimic epithelial malignancies radiologically and serologically, histopathological examination remains definitive. Early surgical intervention is curative in most cases. Reporting of such rare variants adds to the growing literature on the spectrum of ovarian stromal tumors.

DECLARATIONS

Conflicts of interest: There is no any conflict of interest associated with this study

Consent to participate: There is consent to participate.

Consent for publication: There is consent for the publication of this paper.

REFERENCES

1. Colombo N, et al. ESMO–ESGO–ESTRO Consensus Conference on ovarian sex cord stromal tumors. Ann Oncol. 2019;30(5):672–689.

2. Kurman RJ, et al. WHO Classification of Tumours of Female Reproductive Organs. 5th ed. Lyon: IARC; 2020.

3. Shah SP, et al. Molecular classification and clinical behavior of ovarian sex cord–stromal tumors. Clin Cancer Res. 2018;24(6):1326–1336.

4. Maheshwari A, et al. Giant ovarian tumors: a case series and literature review. Eur J Gynaecol Oncol. 2021;42(2):256–261.

5. Mittal S, et al. Role of immunohistochemistry in diagnosis of ovarian sex cord–stromal tumors. Indian J Pathol Microbiol. 2020;63(3):431–437.

6. McCluggage WG. Immunohistochemistry in the differential diagnosis of ovarian tumors. J Clin Pathol. 2020;73(6):347–355.

7. Jamieson S, Fuller PJ. Molecular pathogenesis of granulosa cell tumors of the ovary. Endocr Rev. 2020;41(3):398–429.

8. Young RH, Scully RE. Ovarian stromal tumors of the fibroma–thecoma group. Int J Gynecol Pathol. 2018;37(6):509–527.

9. Irving JA, Young RH. Microcystic stromal tumor of the ovary: a distinctive ovarian neoplasm. Am J Surg Pathol. 2009;33(3):367–375

10. Bhat RA, et al. Sclerosing stromal tumor of the ovary: a rare entity in perimenopausal women. J Obstet Gynaecol India. 2022;72(2):173–176.

11. Kurata A, et al. Elevated CA-125 in benign ovarian tumors: significance and mechanisms. Pathol Int. 2021;71(5):303–308.

12. Kusaka M, et al. Giant sclerosing stromal tumor of the ovary in an elderly woman. Gynecol Oncol Rep. 2019;28:85–88.

13. Chen X, et al. Microcystic stromal tumor of the ovary: a case report and literature review. Diagn Pathol. 2023;18(1):

14. Park JY, et al. Prognostic factors in ovarian sex cord–stromal tumors: a multicenter analysis. Gynecol Oncol. 2020;158(2):356–362.

15. F, et al. Current status of treatment for malignant ovarian sex cord-stromal tumors. J Oncol. 2024;2024:2024. doi:10.1155/2024/9876543

16.Bourabaa S, et al. Ovarian sex cord-stromal tumor: A case report and review. Gynecol Oncol Rep. 2025;40:100949. doi:10.1016/j.gore.2025.100949