A Sclerosing Stromal Tumour of the Ovary in a postmenopausal Female: A Rare Case Report

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Research Article

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Abstract

Sclerosing stromal tumour (SST) is an extremely rare benign ovarian neoplasm, usually seen in young females. We report a case of a 55-year-old female who presented with pelvic pain and irregular bleeding. On imaging, she was suspected of have a borderline malignant tumour of the left ovary and benign teratoma of the right ovary. Postoperatively, diagnosis of SST of left ovary and mature cystic teratoma of right ovary was made after histopathological examination and immunohistochemistry. Our case is unique because SST is a rare entity, and its existence with teratoma has not been reported to date in the literature; and lastly, its occurrence in elderly females is also rarer.

Introduction

Sclerosing stromal tumour (SST) of the ovary is an extremely rare benign neoplasm, and accounts for 2-6% of ovarian sex cord-stromal tumours. They were first described in the literature in 1973 by Chalvardjan and Scully.^{1,2} They are primarily reported in young females in second to third decade and are hormonally inactive. Patients mainly present with pelvic pain, menstrual disturbances, and usually a large pelvic mass with a mean diameter of 10cm.³ We report a rare case of SST occurring in a 55-year female in one ovary and associated mature cystic teratoma in another ovary and discuss its clinical, radiological, and histopathological findings.

Case Report

A 55-year-old female (G6P6) presented with postmenopausal bleeding and dull aching pelvic pain for five months. There was mild tenderness in the left lower abdomen without guarding on physical examination, and her vitals were stable. On pelvic examination, there was left adnexal tenderness. Laboratory investigations, including tumour markers and hormonal levels, were within normal limits except Inhibin-B levels which were mildly elevated (93 pg/ml). There was no significant past medical or family history. The patient was advised to undergo MRI for further evaluation.

MRI of the patient revealed a well-defined multiloculated cystic mass, which measured 8.1x13.8x11.7cm, and appeared T1 hypointense and T2 hyperintense with uniform thin enhancing wall and solid component with no definite invasion into surrounding viscera in the left adnexa. A focal fat-containing lesion (hyperintense on both T1 and T2 and showed suppression on fat-saturated images) measured 3.8 x 3.3 cm with well-defined walls was noted in the right adnexa. Both ovaries were not separately visualized from the respective lesions. A provisional diagnosis of a borderline malignant tumour of ovary was made for the left adnexa based on imaging features. The right adnexal lesion was diagnosed as mature cystic teratoma.

Due to the patient's postmenopausal status and willingness to remove the uterus and bilateral adnexa as a therapeutic procedure, a total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. The resected gross specimen showed a 13x12x7cm large left ovarian mass with

predominantly cystic and foci of small homogenous grey-white solid area (Figure 1) The right-sided ovary measured 3.3x3x2cm, and the cut surface was filled with pultaceous material and few hairs.

A microscopic examination from both cystic and solid areas of the left ovary revealed an encapsulated tumour composed of hypo and hypercellular areas (Figure 2a), forming pseudo lobules at places. Two types of cells were identified, spindle to oval-shaped cells with minimal atypia, elongated nuclei, scant eosinophilic cytoplasm, and the epithelioid cells having round vesicular nuclei with prominent nucleoli and moderate cytoplasm (Figure 2b). Stroma adjacent to tumour cells showed areas of collagen deposition highlighted by Masson's trichrome (Figure 3) and the presence of vessels. No mitosis or necrosis seen. No normal ovarian stroma was identified.

Immunohistochemistry revealed immunoreactivity of tumour cells for Vimentin, (Figure 4a) Inhibin, AR, and focal for SMA. The tumour cells were negative to Desmin, EMA, CD99, and CK (Figure 4b). Therefore, a diagnosis of a sclerosing stromal tumour of the left ovary was rendered. Microscopic examination of the right ovary was suggestive of mature cystic teratoma.

Discussion

Ovarian sex cord-stromal tumours are relatively rare neoplasms derived from sex cord and ovarian stroma. They account for 5-8% of all primary ovarian neoplasms.⁴ This group of neoplasm includes granulosa cell tumours, fibroma, thecoma, Sertoli-Leydig cell tumours, steroid cell tumours, and SSTs, wherein SSTs accounts for only 6% of all sex cord-stromal tumours.^{5,6}

Unlike other ovarian stromal tumours in the fifth to the sixth decade, SSTs are most seen in the second to third decade. Patients usually present with pelvic pain, menstrual irregularities, and nonspecific symptoms related to pelvic mass.³ Our patient presented with pelvic pain and menstrual irregularities.

SSTs are usually hormonally inactive tumours, with few reported cases been associated with raised serum androgen levels.³ Very few cases published in the literature also suggest their association with raised estrogen levels.⁷

Owing to the rarity of these tumours, it is not easy to diagnose them preoperatively based on clinical and radiological findings. Therefore, histopathology with immunohistochemistry plays a pivotal role in diagnosis.

On Ultrasound, SSTs are usually solid cystic multilocular lesions with increased peripheral vascularity. On MRI, SSTs show similar features appearing as heterogenous solid cystic mass (cystic component appearing as T2 hyperintense) with marked early peripheral enhancement and centripetal progression.⁸

Grossly, the appearance of these tumours remains variable and can present as small to large masses with variable solid to cystic cut surface.⁹ Histopathology and immunohistochemistry remain essential in differentiating them from other tumours of the same lineage and giving a definitive diagnosis. On

microscopy, SSTs are essentially composed of cellular areas predominantly forming pseudo lobules and hypocellular areas. The cell population in the cellular areas comprises two types, spindle cells and round to oval cells containing lipid. Marked vascular proliferation is another peculiar feature of this tumour.¹⁰

Few differential diagnoses that need to be ruled out while making a diagnosis of SSTs include other sex cord-stromal tumours like fibroma, thecoma, and ovarian edema in few instances.^{10,11} They can be excluded based on histopathological and immunohistochemical markers.

The microscopic features that favor SSTs over fibroma and thecomas are cellular heterogeneity, prominent vascularity, and pseudo lobular pattern.⁸ Sometimes, the vacuolated cells of SSTs may give a signet ring cell-like appearance, and then the Krukenberg tumour needs to be excluded. In those cases, it is essential to remember that the cells of the Krukenberg tumour show nuclear atypia, mitosis and usually contain mucin rather than lipid. They also lack pseudo lobular patterns and are bilateral.¹¹ In our case, bilateral benign ovarian tumours with different morphology and signet ring-like cells were missing.

Ovarian edema can be differentiated from SSTs by the presence of normal ovarian tissue in edematous areas and lack of heterogeneity.¹¹

On immunohistochemistry, SSTs shows immunoreactivity for smooth muscle actin (SMA), vimentin, and inhibin, suggesting its stromal origin and are usually negative for S-100 protein and epithelial markers.^{11,12} CD34 highlights the prominent branching vascular channels in SSTs and differentiates them from fibroma and thecoma.¹³ In this case, the tumour cells showed positivity for inhibin and SMA but were negative for epithelial markers and S-100 protein.

SSTs are benign tumours with a good prognosis and are treated by surgery alone with no known local or distant recurrences except for one case that has been reported as low-grade malignant in the literature.^{9,14} In our case, as the patient was postmenopausal, the uterus with bilateral adnexa was removed, and the postoperative period of the patient remained uneventful.

Mature cystic teratomas of the ovary are the most common benign germ cell tumour of the ovary and are derived from one or more of the three germ cell layers. Most of these tumours occur in reproductive age group females, usually below 40 years of age. Malignant transformation is a rare complication and occurs in 1-3% cases, more commonly seen in postmenopausal women.^{15,16}

The association of benign teratoma had been reported with few neoplasms like mucinous cystadenoma, Brenner tumour, and granulosa cell tumour, but this too is relatively infrequent and the pathogenesis behind them occurring together remains unclear.¹⁷

After extensive literature search and scrutiny, only one case of SST with bilateral teratoma was published in the literature by valentine et al. However, it was a benign mature teratoma and SST of the same ovary and immature teratoma of the other ovary in the published report.¹⁷ Our case is unusual and unique in the way that SST had been diagnosed in one ovary and benign teratoma in the other ovary.

Conclusion

SSTs are benign rare ovarian tumours of young females. It is not easy to diagnose them clinic radiologically, and histopathology remains the definitive tool for diagnosis. In our case, we highlight the occurrence of SST of one ovary in a 55-year-old elderly female and its previously unreported association with mature cystic teratoma of another ovary: further underlining its uniqueness.

Declarations

- i. Funding: NIL
- ii. Conflicts of interest/Competing interests: NIL
- iii. Ethics approval: Waived
- iv. Consent to participate: Patient has given consent to participate
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- vii. Code availability (software application or custom code): NA
- viii. Authors' contributions

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Planning	✓	✓	
Conduct		✓	✓
Reporting	✓	✓	
Concepts & Design		✓	✓
Data acquisition		✓	✓
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Figures

Figure 1

Cut surface of left ovarian solid cystic mass, the solid nodule is grey, white.

Figure 2

a, b: Tumour cells are showing hypercellular and hypocellular area. [HEx10; x40]

Figure 3

Masson trichrome highlight collagen and fibroblast. [x10]

Figure 4

a: Tumour cells are immunoreactive to Vimentin. [x20]

b: Tumour cells are negative to Cytokeratin. [x20]

Supplementary Files

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