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

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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 having a borderline malignant tumour of the left ovary and benign teratoma of the right ovary. Postoperatively, diagnosis of SST of the left ovary and mature cystic teratoma of the 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.

Keywords Stromal tumour · Postmenopausal female · Elderly female · Benign ovarian neoplasm

Introduction
Sclerosing stromal tumour (SST) of the ovary is an extremely rare benign ovarian 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 the second to the 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 10 cm [3]. We report a rare case of SST occurring in a 55-year-old 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 5 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 like CA-125 and CEA, were within normal limits except inhibin-B levels which were mildly elevated (93 pg/ml). There was no significant past medical or family history. Oestrogen or progesterone levels were not done pre- or postoperatively. Calcitonin, CD34, and α-glutathione S-transferase (αGST) markers were not evaluated due to the unavailability of these markers at our institute. The patient was advised to undergo MRI for further evaluation.

CT findings (Fig. 1a) revealed a well-defined cystic mass in the left adnexa measuring 8.1 × 13.8 × 11.7 cm with uniform wall enhancement and solid component without any definitive invasion into surrounding viscera. A focal fat density lesion with well-defined walls measuring 3.8 × 3.3 cm was noted in the right adnexa.

MRI of the patient revealed a well-defined multiloculated cystic mass, which measured 8.1 × 13.8 × 11.7 cm, 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.

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A focal fat-containing lesion (hyperintense on both T1 and T2 and showed suppression on fat-saturated images) measured 3.8 × 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 the ovary was made for the left adnexa based on imaging features. The right adnexal lesion was diagnosed as mature cystic teratoma. In view of postmenopausal bleeding, an endometrial biopsy was done which showed features of the atrophic polyp.

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. Intraoperative findings showed a 15 × 12 × 1 cm cystic swelling arising from the left ovary with 500 ml of peritoneal fluid collection. The resected gross specimen showed a 13 × 12 × 7 cm large left ovarian mass with predominantly cystic and foci of small homogenous grey-white solid area (Fig. 1b) The right-sided ovary measured 3.3 × 3 × 2 cm, 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 (Fig. 2a), forming pseudolobules 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 (Fig. 2b). Stroma adjacent to tumour cells showed areas of collagen deposition highlighted by Masson’s trichrome (Fig. 3) and the presence of vessels. No mitosis or necrosis seen. No normal ovarian stroma was identified. Endometrium showed features of an atrophic polyp.

Immunohistochemistry revealed immunoreactivity of tumour cells for vimentin (Fig. 4a), inhibin, androgen receptor (AR), and focal for smooth muscle actin (SMA). The tumour cells were negative to desmin, epithelial membrane antigen (EMA), CD99, and cytokeratin (CK) (Fig. 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. On follow up of the patient for a period of 2 years, there was no evidence of recurrent or residual lesion.

**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 account 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 oestrogen 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 surfaces [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 pseudolobules and hypocellular areas. The cell population in the cellular areas

Fig. 3  Masson trichrome highlights collagen and fibroblast. [MT × 10]

Fig. 4  a Tumour cells are immunoreactive to vimentin. [IHC × 20]. b Tumour cells are negative to cytokeratin. [IHC × 20]
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 oedema in a few instances [10, 11]. They can be excluded based on histopathological and immunohistochemical markers.

The microscopic features that favour SSTs over fibroma and thecomas are cellular heterogeneity, prominent vascularity, and pseudolobular pattern [8]. Sometimes, the vacuolated cells of SSTs may give a signet ring cell-like appearance, and then the possibility of 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 pseudolobular patterns and are bilateral [11]. In our case, bilateral benign ovarian tumours with different morphology were there and signet ring-like cells were missing. Alcian blue in the combination with Periodic acid-Schiff (AB-PAS) at pH 2.5 highlights the cytoplasmic mucin in the signet ring cells of Krukenberg tumour whereas SSTs are non-reactive for AB-PAS stain.

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

On immunohistochemistry, SSTs show immunoreactivity for 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% of 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 clinically, 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.

**Author Contribution** Planning — Pallavi Saraf, Jyotsna Naresh Bharti. Conduct — Jyotsna Naresh Bharti, Charu Sharma. Reporting — Pallavi Saraf, Jyotsna Naresh Bharti. Concepts and design — Jyotsna Naresh Bharti, Charu Sharma. Data acquisition — Jyotsna Naresh Bharti, Charu Sharma. Data interpretation — Pallavi Saraf, Jyotsna Naresh Bharti. Manuscript preparation, editing and review — Pallavi Saraf, Jyotsna Naresh Bharti.

**Data Availability** All relevant data and material is available with the corresponding author.

**Code Availability** Not applicable.

**Declarations**

**Ethics Approval** For this type of study, formal consent is not required and was performed under a waiver of informed consent by the Institutional Review Board, AIIMS Jodhpur.

**Consent to Participate** The authors certify that they have obtained all appropriate patient written consent format. The patient has given her consent for their images and other clinical information to be reported in the form. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity.

**Conflict of Interest** The authors declare no competing interests.

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