CASE REPORT

Ovarian Sclerosing Stromal Tumour: A Rare Stromal Neoplasm With Distinctive Histopathology

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ABSTRACT

Sclerosing stromal tumour (SST) represents an uncommon benign neoplasm of ovarian stromal origin that typically affects women in their second and third decades. We report a case of a 26-year-old female who presented with lower abdominal discomfort, irregular menstruation, and a palpable mass in the pelvis. Laboratory assessment showed normal levels of tumour markers, including CA-125 and alpha-fetoprotein. Abdominal and pelvic computed tomography (CT) imaging demonstrated a complex solid-cystic lesion with calcified areas originating from the right ovary, measuring 4.3 × 3.9 × 4.8 cm, without radiological indications of malignancy. Despite benign imaging characteristics, the clinical presentation necessitated surgical intervention to rule out malignancy, leading to a right salpingo-oophorectomy. Paucity of reported cases in Southeast Asian region, may contribute to this uncertainty that could reflects the inadequate systematic reporting, potential underdiagnosis, or misclassification of SSTs as other stromal neoplasms. Thus, the reporting of individual cases, including those with characteristic presentations, remains especially important for enhancing our knowledge of population-specific patterns and strengthening diagnostic recognition within the medical community. The definitive SST diagnosis resulted from comprehensive multidisciplinary assessment incorporating clinical, radiological and histopathological findings. This case underscores the necessity of thorough diagnostic evaluation to distinguish SST from other ovarian lesions, particularly in young women with adnexal masses.
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INTRODUCTION

Sclerosing stromal tumour (SST) signifies an uncommon benign neoplasm originating from the sex cord-stromal elements of the ovary, initially described by Chalvardjian and Scully in 1973 [1]. SST constitutes roughly 5% of ovarian stromal tumours [2]. This condition primarily affects young women and typically manifests as a unilateral ovarian mass. The clinical presentation commonly includes pelvic pain and menstrual abnormalities, despite the tumour generally lacking hormonal manifestation. SST characteristically appears as a well-demarcated solid-cystic lesion on imaging studies, that frequently raises suspicions of malignancy preoperatively. On that account, definitive diagnosis depends on histopathological evaluation. To date, only a single case of recurrence has been reported in the literature, which was linked to capsular rupture, necrosis, and increased mitotic activity. Otherwise, complete surgical excision continues to be the preferred treatment approach.

Within the Asian region, Korea, China, and India have contributed most of the published case reports of sclerosing stromal tumours. For Southeast Asia specifically, the literature reveals no reported case from Malaysia, while a recent publication from Indonesia described an SST case characterized by masculinising symptoms. Several factors may account for the paucity of reported cases, including inadequate systematic reporting, potential underdiagnosis, or misclassification of SSTs as other stromal neoplasms. Hence, case documentation of even conventionally presenting SSTs becomes particularly meaningful in advancing our understanding of population-specific features and fostering greater diagnostic recognition among healthcare providers. This report describes the clinical features, distinctive morphological and immunohistochemical characteristics as well as imaging characteristics of SST

and emphasizes its differentiation from other similarly presenting ovarian neoplasms.

CASE REPORT

A 26-year-old nulliparous woman sought medical attention for persistent pelvic discomfort and menstrual irregularities spanning several months. Physical examination identified a firm, non-painful mass that could be palpated in the right lower abdominal quadrant. Laboratory assessment revealed tumour markers within normal ranges, including alpha-fetoprotein (α-FP), cancer antigen 125 (CA-125), and beta-human chorionic gonadotropin (β-hCG). Computed tomography (CT) imaging of the abdomen and pelvis revealed a heterogeneously enhancing right adnexal lesion measuring approximately 43 × 39 × 48 mm, characterized by central hypodense regions and calcifications (Fig. 1).

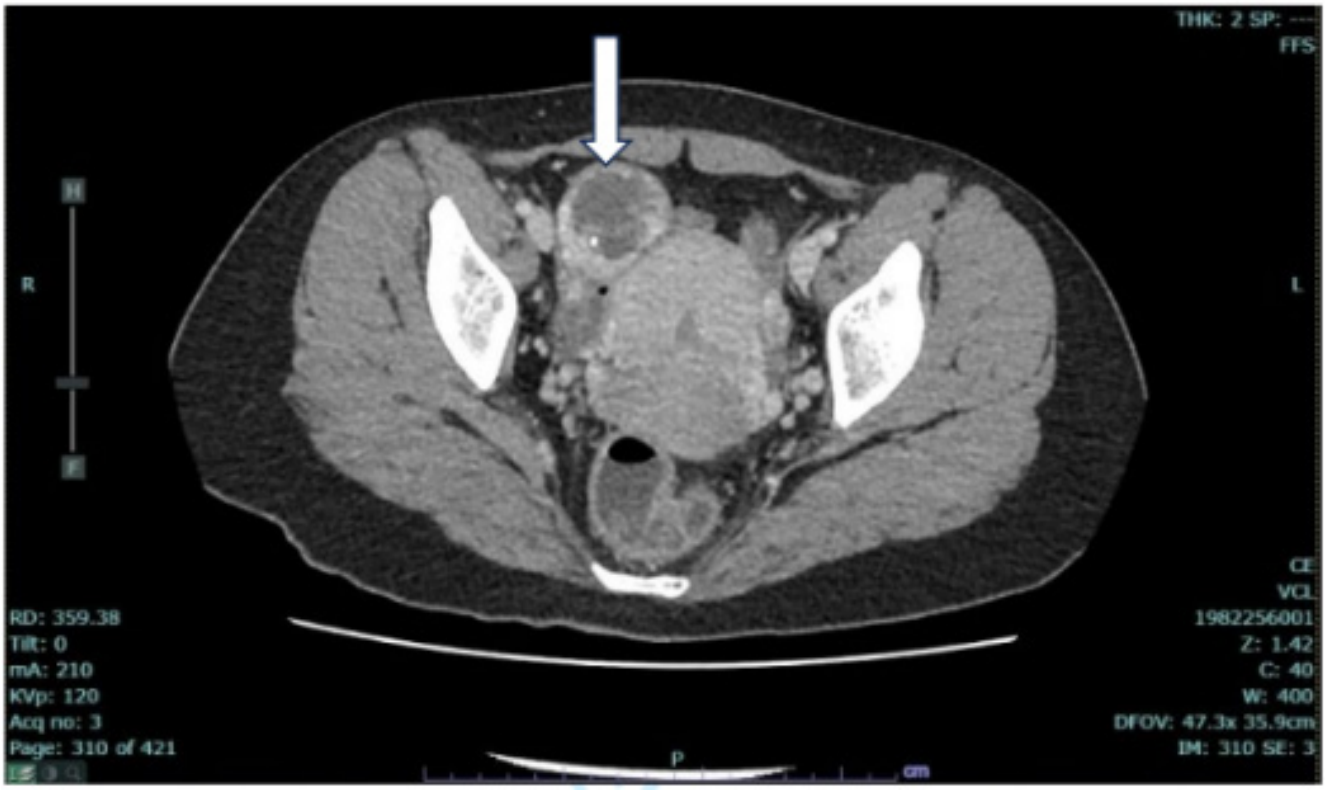

Fig. 1: CT abdomen and pelvis. Right ovarian heterogeneously enhancing mass with central hypodensity and calcification, size 4.3x3.9x4.8cm (APxWxCC) (arrow) with presence of intracystic irregular frond like papillary projections with no fat component.

The patient underwent an open surgical procedure for right salpingo-oophorectomy. During surgery, a well-encapsulated mass measuring 40 mm × 40 mm was identified in the right ovary. The contralateral ovary and uterus appeared normal on visual inspection, and no peritoneal fluid accumulation was detected. Macroscopic assessment of the resected ovary showed an intact, smooth external capsule. Cross-sectional examination revealed a predominantly solid tumour with firm consistency and whitish-yellow coloration, interrupted by multiple cystic spaces that measures from 5 to 15 mm in cyst diameter, some containing clear transparent fluid.

Microscopic examination demonstrated a characteristic pseudolobular pattern with alternating hypercellular and hypocellular regions (Fig. 2A). The hypercellular areas consisted of dual populations of benign-appearing epithelioid and spindle cells (Fig. 2B). The hypocellular zones featured dense collagenous stroma with focal oedematous and myxoid changes. Distinctive hemangiopericytoma-like vascular patterns were observed throughout the specimen. Immunohistochemical analysis showed diffuse positivity for vimentin and β-catenin (Fig. 2C-D), focal calretinin expression, and patchy, weak α-inhibin reactivity (Fig. 2E-F). The overall findings confirmed the diagnosis of benign sclerosing stromal tumour of the ovary.

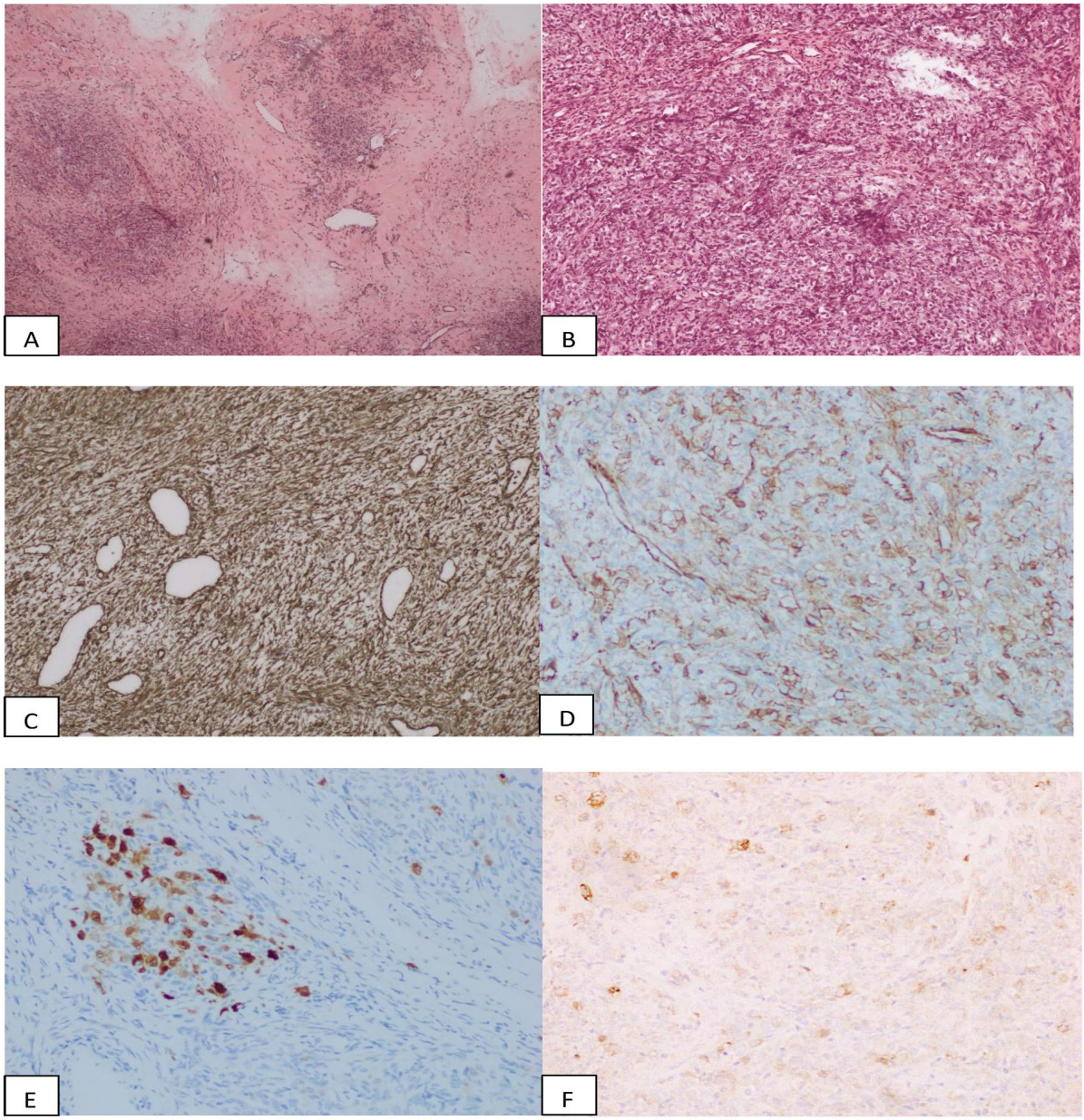

Fig. 2: A) Pseudolobular appearance imparted by alternating cellular and hypocellular areas (H&E staining, x40). B) Cellular nodules consist of two populations of bland epithelioid and spindled cells (H&E staining, x100). C,D) Diffuse cytoplasmic staining for Vimentin and β catenin (immunohistochemistry, x100). E,F) Focal positive (cytoplasmic and nuclear) Calretinin staining and weakly positive patchy granular cytoplasmic staining of α-inhibin (immunohistochemistry, x100).

DISCUSSION

Sclerosing stromal tumour (SST) of the ovary characterises an uncommon benign neoplasm originating from sex cord-stromal tissue, constituting less than 1% of all ovarian tumours with roughly 200 cases reported in medical literature [2]. This tumour distinctively affect younger females, with approximately 80 percent of diagnoses occurring in women in their second and third decades [3,4]. This age distribution contrasts with other ovarian stromal neoplasms, which typically manifest in women aged 40-60 years.

Clinical presentation commonly includes nonspecific manifestations such as pelvic pain, menstrual abnormalities, or detection of an adnexal mass. While most SSTs remain hormonally inactive, a small percentage may produce endocrine effects including mild virilisation. An earlier publication described a case of SST presenting with masculinizing features over a seven-year period, occurring in conjunction with Meig's syndrome [3]. Peritoneal fluid accumulation rarely accompanies SST, and was absent in the current case.

When imaging identifies an ovarian mass in a young female patient, germ cell tumours constitute a significant differential diagnosis consideration. As such, tumour markers including α-FP, β-hCG, and CA-125 may be assessed as part of the diagnostic approach. These markers typically do not contribute significantly to the initial diagnostic workup but serve primarily as monitoring tools for disease recurrence detection. Baseline tumour marker levels are often obtained when ovarian malignancy is suspected for future comparison. While tumour markers are generally normal in sclerosing stromal tumours (SSTs), exceptions exist. Occasional reports demonstrated elevated CA-125 levels in a patient with histologically confirmed SST, highlighting that these benign tumours can occasionally present with abnormal tumour marker values [3]. This finding underscores the limitation of relying solely on tumour markers for diagnosis and emphasizes the importance of histopathological confirmation for definitive diagnosis.

Among the ovarian stromal neoplasms, SST exhibits distinctive histological features. The tumour demonstrates a characteristic pseudolobular architecture with alternating regions of high and low cellularity. Within the cellular lobules, a heterogeneous population of spindle cells and rounded epithelioid cells containing lipid and lutein material can be observed, along with conspicuous branching vasculature resembling hemangiopericytoma-like patterns. Dense collagenous stroma separates these cellular components. Identifying this specific morphological signature is essential for establishing an accurate diagnosis [1,3-5]. Current theory suggests that these tumours arise from the stromal components within the ovarian cortex, though the exact pathogenic mechanisms have yet to be fully clarified.

When considering SST, it must be differentiated from other sex cord-stromal neoplasms including fibromas, thecomas, and granulosa cell tumours as in Table 1 [1,3,5]. While imaging techniques can aid in the diagnosis, conclusive identification requires histopathological evaluation. Radiographically, SST typically presents as a circumscribed mass with both solid and cystic components, potentially resembling malignant ovarian lesions. However, its macroscopic and microscopic features facilitate distinction from other tumours. Fibromas and thecomas, for instance, lack the characteristic pseudolobular arrangement and predominantly affect older women. These entities usually manifest with generalized symptoms including pelvic discomfort, abdominal enlargement, or urinary symptoms. Endocrine activity is uncommon in these tumours, though larger fibromas may present with ascites. Thecomas frequently produce oestrogen, potentially causing postmenopausal uterine bleeding, endometrial proliferative changes, or in some cases, endometrial malignancy.

Table I: Clinical and histological characteristics of SSTs and its differentials of sex cord stromal tumour (SCST) of ovary.

Clinical and histological characteristics	Sclerosing stromal tumour	Granulosa cell tumour (AGCT)	Fibroma	Thecoma
Epidemiology	Uncommon, young women and girls, ~5% of all SCST	1% of all ovarian tumours	4% of all ovarian neoplasms (most common ovarian stromal tumour)	Uncommon
Age	20-30s, (Mean age 29)	Any age, perimenopausal	Any age; middle age (average age 48 years)	Postmenopausal (Mean age 59)
Symptoms	-AUB, mass effect, incidental findings. -Hormonal symptoms uncommon.	-Abdominal pain. -Oestrogenic manifestations eg: uterine bleeding. -Occasional virilisation	-Mass effect. -Hormonal manifestations.	-Mass effect. -Hormonal manifestations (estrogenic or less commonly androgenic)
Tumour markers	-	elevated serum β-inhibin	-	-
Other features	Rarely associated with Meigs syndrome	Concurrent endometrial hyperplasia	Associated with Gorlin syndrome, Meigs syndrome	Associated with endometrial proliferations
Macroscopic	-Unilateral, well circumscribed, solid yellow to white - 1.5 to 19 cm (mean 11 cm) -Central oedema and cyst formation.	-Unilateral, solid and cystic, soft and tan to yellow -Average size of 10 cm -Haemorrhage is common.	-Unilateral or bilateral, smooth capsule, hard, chalky, white or yellowish-white or tan. -Oedema, cystic degeneration, haemorrhage or necrosis may be present.	-Unilateral or bilateral, solid, yellowish-tan or focally white -Size 5–10 cm in diameter or larger -Cystic, haemorrhagic, or necrotic areas occasionally seen.
Microscopic	Pseudolobular appearance, cellular nodules of epithelioid and spindled cells separated by hypocellular, oedematous, collagenous, or occasionally myxoid stroma, low mitotic activity.	Variety of architectural patterns, (cords, trabeculae, insular or microfollicular Call–Exner bodies pattern). Spindled, round to oval nuclei, irregular nuclear membrane, nuclear grooves, and scant cytoplasm. Low mitotic activity.	Intersecting fascicles of cells with bland, spindled to ovoid nuclei and scant cytoplasm within a variably collagenous stroma. Mitoses are uncommon. Haemorrhage and infarct-type necrosis may occur.	Diffuse, lobulated/nested uniform cells with ovoid to round, sometimes with small nucleoli. Nuclear grooves can be seen. Mitotic activity is either absent or minimal. May have hyaline plaques, sometimes forming keloid-like sclerosis.
Special stains and IHC	Immunoreactive to sex cord markers, inhibin and calretinin, but negative for CK and EMA. TFE3 positive in a subset of tumours. β catenin- positive (nuclear), Melan A -positive CD99 - Weak WT1- usually negative	Reticulin stain - highlights reticulin fibres around tumour nests. Immunoreactive to FOXL2, calretinin, inhibin (vary in distribution and intensity), SF1, ER, panCK, CD99, WT1 and Vimentin are frequently positive. PAX8, CK7, and EMA are typically negative.	Reticulin stain – highlight individual cells. May be immunoreactive for inhibin (focal and weak), calretinin (focal and weak), WT1, FOXL2, CD56, SF1, Vimentin and hormone receptors.	Reticulin stain - usually surrounds individual cells. Typically positive for inhibin, calretinin, WT1, SF1, FOXL2, Vimentin.
Molecular	Subset of trisomy 12 Some with recurrent FHL2-GLI2 fusion genes.	FOXL2 point mutation.	NA	Occasional FOXL2 mutations (unclear if represent misdiagnosed AGCT).

Granulosa cell tumours (GCT) are another key differential consideration, typically characterized by hyper-oestrogenic states. In younger patients, clinical manifestations often include irregular uterine bleeding, endometrial abnormalities, or precocious sexual development. In rare instances, these tumours may secrete androgens, leading to virilising symptoms. Morphologically, they commonly present as large, unilateral masses with mixed solid and cystic components, occasionally complicated by torsion, rupture, or haemoperitoneum. Juvenile granulosa cell tumour (JGCT) mostly occurs in adolescent females. Cysts of varying sizes with microfollicles containing eosinophilic secretions are the differentiating features [3].

The radiological differentiation of SSTs presents significant challenges due to shared imaging characteristics with both malignant and benign ovarian lesions. SST typically demonstrates a mixed solid-cystic appearance with heterogenous enhancement on ultrasound and CT scan. In comparison, fibromas characteristically appear as homogenous hypoechoic masses with posterior acoustic shadowing, while thecomas present as well-defined hypoechoic lesions [2]. Granulosa cell tumours frequently manifest as complex masses containing both solid and fluid-filled regions. Despite these similarities in appearance, SST often exhibits characteristic increased vascularity, detectable through colour Doppler ultrasound, CT, or Magnetic Resonance Imaging (MRI). This distinctive vascular pattern has been proposed as a valuable diagnostic indicator for distinguishing SST from other ovarian tumours. Another distinguishing feature that is useful to characterise SST is avid contrast enhancement from the periphery to the center on MRI. The centripetal enhancement pattern has been described as hemangioma-like [2].

In cases where diagnostic uncertainty persists, immunohistochemistry offers further clarification. SST characteristically exhibits positive immunoreactivity for stromal markers including α-inhibin, calretinin, and vimentin, while consistently showing negative expression for epithelial markers such as cytokeratin (CK) and epithelial membrane antigen (EMA) [3,4]. Further immunohistochemical distinction can be obtained through assessment of β-catenin, Melan-A, CD34, CD99, and WT-1, which assist in differentiating SST from related sex cord-stromal neoplasms. The immunoprofile of SSTs includes positivity for β-catenin and Melan-A, with mostly weak or negative staining for CD99 and WT-1. GCT may be positive for β-catenin but is mostly FOXL2 immunoreactive. Fibroma and thecoma are less likely to show β-catenin positivity. These differentials of SST listed in Table 1 show positive WT-1 expression. CD34 immunostaining accentuates the characteristic prominent vascular architecture seen in SST. Of significant interest, recent work by Zhao and colleagues (2023) has proposed the nuclear transcription factor TFE3 as a potentially selective immunohistochemical marker for SST, providing particular diagnostic value in challenging cases or those with predominantly cystic morphology [4].

CONCLUSION

In summary, SST embodies an uncommon but well-characterized ovarian neoplasm that warrants consideration when evaluating adnexal masses in women during their teens and twenties. Because of its nonspecific clinical manifestations and imaging characteristics that overlap with other ovarian lesions, conclusive preoperative identification remains difficult and depends substantially on histological and immunohistochemical assessment. Consequently, it should be included in the differential diagnoses, when encountering unilateral, mixed solid-cystic ovarian masses in young female patients. Awareness of this entity and recognition of its characteristic morphological and vascular patterns can enhance diagnostic accuracy, enabling appropriate conservative surgical approaches, such as cystectomy, which typically provide adequate treatment while maintaining reproductive potential in most patients.

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