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Case Report

# Cystic sclerosing stromal tumor of the ovary: A case report and review of literature

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## ABSTRACT

Sclerosing stromal tumor (SST) of the ovary is an uncommon benign ovarian neoplasm of sex cord stromal origin with distinct clinical and radiological features. We describe a cystic SST in a 35-year-old female who presented with right lower abdominal pain. Computerized tomography scan revealed a complex ovarian cyst. Right-sided Salpingo-oophorectomy was done with an intraoperative frozen section, which ruled out malignancy. Histopathology showed features consistent with SST ovary. In this report, we discuss the differential diagnosis and role of histopathology in confirming the benign nature of the neoplasm, so a conservative surgery can be performed.

KEY WORDS: Abdominal pain, cellular, cystic, luteinization, sclerosing stromal tumor

## INTRODUCTION

Sclerosing stromal tumor (SST) of the ovary is an uncommon benign ovarian neoplasm with distinct clinical, pathological, and radiological features which distinguish them from other ovarian sex cord stromal tumors [1,2].

They usually affect young individuals and manifest as menstrual irregularity and/or pelvic pain [3].

We present a case of cystic SST of the right ovary encountered in Kasturba Medical College, Manipal.

## CASE REPORT

A 35-year-old woman reported to the Department of Surgical Oncology with pain in the right lower abdomen of 15 days duration. A physical examination and medical history of the patient was unremarkable. Serum markers including CA-125, alpha-fetoprotein, Serum β-human chorionic gonadotropin were within normal reference range with values 16.8 U/ml, 4.9 ng/ml, and 0.6 IU/ml, respectively. No virilization or menstrual abnormalities were identified.

Computerized tomography (CT) of the abdomen showed a complex right ovarian cyst. Right-sided Salpingo-oophorectomy was done, and the intraoperative frozen section was performed which ruled out malignancy.

Grossly, an ovarian cyst measuring 12.5 cm × 6 cm × 1 cm with a smooth and glistening external surface was identified attached to a fallopian tube. On cut section, the cyst was uniloculated with solid gray areas with rounded ill-formed excrescences [Figure 1].

Microscopy of the solid areas revealed a lobulated ovarian neoplasm composed of cellular nodules with alternating edematous stroma [Figure 2]. The cellular areas were composed of an admixture of plump spindle-shaped cells with 4 mitosis/10 hpf in active regions and round to ovoid cells with moderate to abundant, pale to eosinophilic cytoplasm focally forming nests and islands with the intervening paucicellular zones showing marked edema with proliferating vessels and spindle-shaped stromal cells, focal areas showing stromal luteinization with the latter abutting onto the periphery of the cellular nodules was also seen [Figures 3 and 4]. Periphery also showed few residual corpus albicans. The fallopian tube showed normal morphology.

Immunohistochemistry revealed patchy positivity for inhibin, calretinin, and smooth muscle actin in the tumor cells [Figures 5-7].

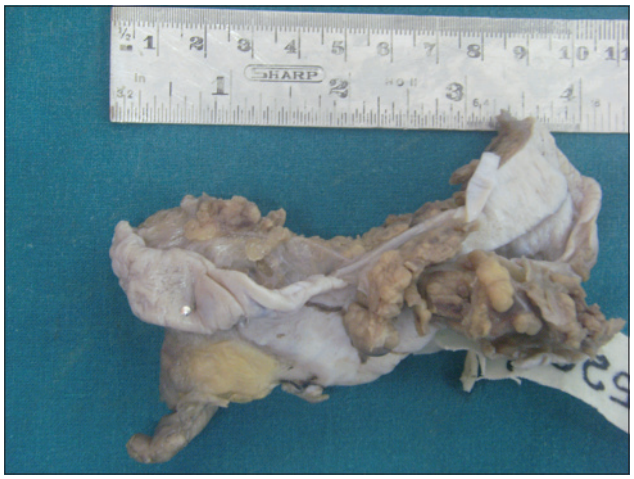

**Figure 1:** Tiny broad papillary excrescences in a predominantly cystic sclerosing stromal tumor of ovary

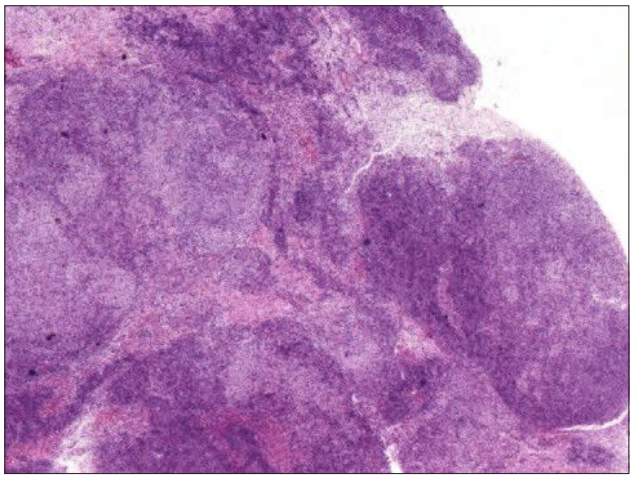

**Figure 2:** Sclerosing stromal tumor of ovary showing hypocellular and hypercellular lobular arrangement (H&E, ×40)

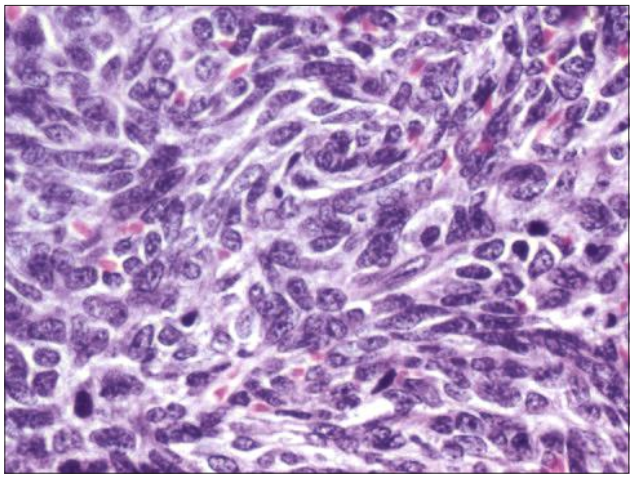

**Figure 3:** Cellular foci of sclerosing stromal tumor of ovary admixture of spindle and round to ovoid cells (H&E, ×400)

A diagnosis of cystic SST of the right ovary was rendered.

The patient has been on regular follow-up for 1 year and is asymptomatic.

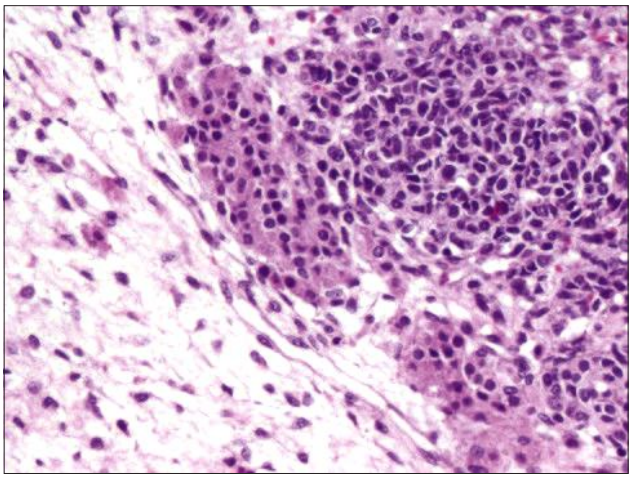

**Figure 4:** Sclerosing stromal tumor of ovary showing focal luteinization (H&E, ×100)

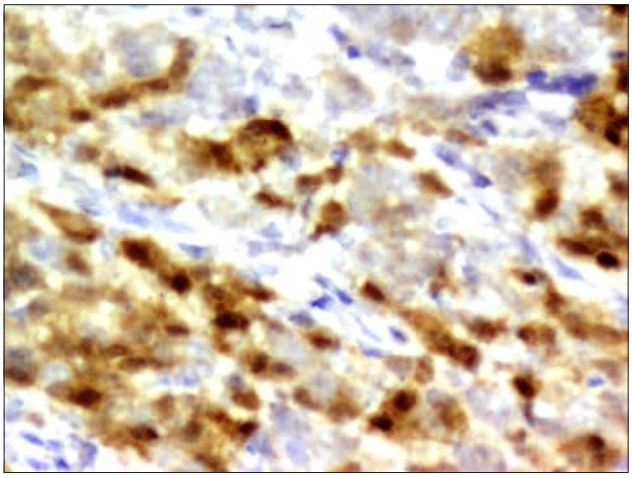

**Figure 5:** Patchy positivity of inhibin (H&E, ×400)

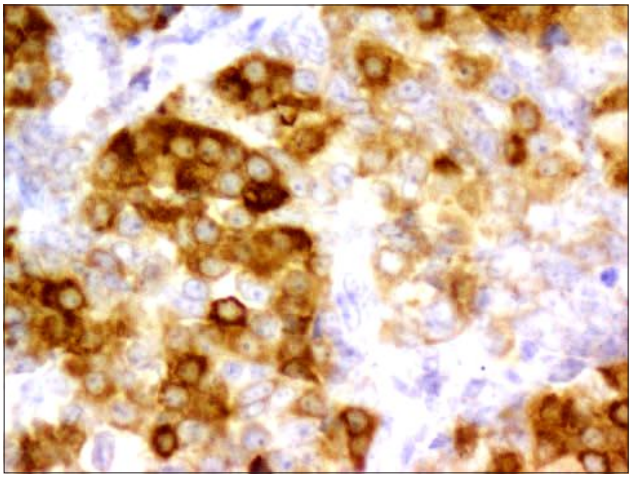

**Figure 6:** Patchy positivity of calretinin (H&E, ×400)

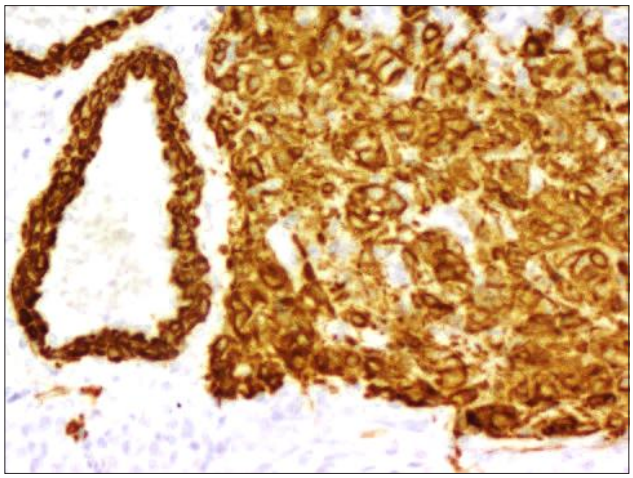

**Figure 7:** Patchy positivity of Smooth muscle actin (H&E, ×200)

## DISCUSSION

Ovarian "SST" is a benign and rare tumor of sex cord stromal origin, which was first described and defined in 1973 by Chalvaridjan and Scully [1,2].

SSTs usually affect individuals in the 2nd and 3rd decades of life. However, pediatric cases have been reported [4,5]. Patients may be asymptomatic or present with menstrual disturbances such as polymenorhea, metrorrhagia, dysmenorrhea, and amenorrhea. Infertility, hirsutism, abdominal mass, abdominal pain, and ascites may also be present [1,3,6]. These tumors are usually hormonally inactive and may also be associated with elevated levels of CA-125 [1,7,8]. In the discussed case, the patient presented with right abdominal pain and normal serum levels of CA-125. No hormonal activity or virilization was observed.

On radiology, SSTs appear as a well-defined mass with increased peripheral enhancement on CT [9]. However, preoperative diagnosis based on clinical and radiological findings alone is difficult [7]. In our case, a complex ovarian cyst was described on CT and no comment on the tumor type was made. The final diagnosis was thus based on the characteristic histopathological features with ancillary immunohistochemical (IHC) studies.

Macroscopically, SSTs range from 1 cm to 31 cm in diameter and usually are unilateral [1,7]. They may be solid or cystic, composed of single large or multiple small mucoid or serous fluid filled cysts. The demarcation between the tumor and the adjacent ovarian stroma may be indistinct [1,2]. Microscopically, on low power examination, the tumor is composed of cellular and hypocellular areas imparting a pseudo lobular pattern. On higher magnification, the cellular areas are composed of cells with varied size and shape arranged haphazardly containing pronounced sclerosis and prominent vascular network arranged in a hemangiopericytomatous pattern. The cells are mostly round or spindle in shape with a vesicular nucleus, prominent nucleolus, sometimes showing luteinization and clear or vacuolated cytoplasm, occasionaly displacing the nuclei to the periphery giving it a signet ring cell appearance. Mitoses are less frequent [1,2,6]. IHC, SSTs show diffuse positivity with Vimentin and progesterone receptor; the spindle-shaped cells show strong positivity for SMA (smooth muscle actin) and desmin. The cells with vacuolized cytoplasm stain positive for inhibin and calretinin. SSTs do not stain for keratin, CD10 and estrogen receptor [1,3,10]. These IHC properties suggest their stromal origin.

The common differential diagnosis of SSTs includes fibroma/the coma, massive edema of ovary and krukenberg tumor. The distinct clinical, histopathological features and IHC characteristics can be helpful in differentiating these entities.

Younger age of presentation, cellular heterogeneity, prominent vascularity and absence of hyalinized plaques along with immunoreactivity to CD34 and alpha glutathione S transferase favors a diagnosis of SST over fibroma/the coma [11]. The preservation of the ovarian tissue within the diffusely edematous stroma in contrast to the focal edema in SSTs, and the lack of cellular heterogeneity distinguish massive edema of the ovary from SSTs. Sometimes SSTs with a predominant signet ring like cells may pose diagnostic dilemma with Krukenberg tumor. They can be differentiated from the latter by the lack of mucicarmine stain and cytokeratin [1,3,7]. Further, history of primary gastrointestinal malignancy is usually present.

In the presented cases, the characteristic histopathological findings aided by the IHC positivity for inhibin, SMA and vimentin helped in confirming a diagnosis of SST.

In conclusion, SSTs which have been reported with such rarity should be considered in young women with the above discussed clinical and radiological findings. In predominantly cystic SSTs, the solid foci will show the characteristic morphologic features. In addition to histopathological evaluation, IHC analysis is necessary for a definite diagnosis. Salphingo-oophorectomy is the treatment of choice, and no recurrences have been reported until date with complete abatement of the symptoms [3,7,12].

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