

Sclerosing Stromal Tumor of Ovary in Adolescence: A Rare Case Report and Brief Review of Literature.

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Abstract

Ovarian sex cord-stromal tumors are relatively infrequent neoplasms that account for approximately 8% of all primary ovarian neoplasms. Due to rarity of this neoplasm, it may be difficult to determine the tumor type preoperatively on radiological findings. Sex cord stromal tumour is distinctive from other sex cord stromal tumors because of its unusual clinical presentation at an early age and lack of hormonal manifestations. We present this uncommon case in a 17 years old female highlighting the importance of histopathology examination and immunohistochemistry (IHC) in the diagnosis of this benign ovarian neoplasm.

Key Words: Ovarian tumor, sclerosing stromal tumor, Pseudo lobular pattern,

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I. Introduction

Sclerosing stromal tumors (SSTs) of the ovary have been described as a pathologically and clinically distinct subgroup within the thecoma-fibroma spectrum of benign ovarian sex cord-stromal tumors. It is a rare ovarian disease with prevalence of 1.5-6% of ovarian stromal tumors occurring predominantly in young women [1]. Fewer than 100 cases with SSTs are described in the literature since its first description in 1973 by Chalvardijan and Scully, indicating the rarity of this entity[2,3]. It is distinguished from other ovarian tumors by the production of collagen and a pseudolobular pattern with cellular areas separated by edematous and collagenous areas. Histopathological and immunohistochemical (IHC) examinations confirm the diagnosis[1]. These tumors occur predominantly in the second and third decade of life[4].

We present this uncommon case of SST in a 17 years old female highlighting the importance of histopathology examination and IHC in the diagnosis of this benign ovarian neoplasm.

II. Case Report

A 17-year-old female presented with menometrorrhagia and lower abdominal pain since 4 years. History of hypothyroidism was present 3 years back for which she took treatment for 4 months. Similar complaints were present in her paternal aunt.

On physical examination, a firm mass in the hypogastric region was noted. Pelvic ultrasonography showed a right adnexal, large heterogenous mass measuring 10x7 cm with minimal internal vascularity and mass is not entirely separated from right ovary. Left ovary was not visualised with uterus and endometrium. Magnetic resonance imaging (MRI) pelvis showed solid appearing right ovarian mass measuring 9.2x6.2x5 cm, medially extending into pouch of douglas and abutting right posterolateral uterine wall, sign of neoplastic origin.(Figure-1) Changes of polycystic ovarian disease(PCOD) were seen in left ovary.(Figure-2)



Figure 1: MRI revealed solid appearing right ovarian mass measuring 9.2x6.2x 5cm medially extending to pouch of douglas abutting right posterolateral uterine wall.

Routine hematological and biochemical investigations were within normal limits. Tumor markers Cancer antigen 125(CA 125), α -fetoprotein and β -human chorionic gonadotrophin (β -hCG) were also within normal limits.

Unilateral salpingo-oophorectomy was conducted and excised specimen of right adnexal mass was sent for histopathology examination.

The gross examination of the resected specimen showed an encapsulated, whitish, globular mass measuring 6x5x2.5 cm. The external surface was smooth and partially cut open. Cut surface was grey white to yellowish, solid with gelatinous areas and multiple cystic spaces were appreciated (Figure3, 4). No hemorrhage or necrosis was noted.

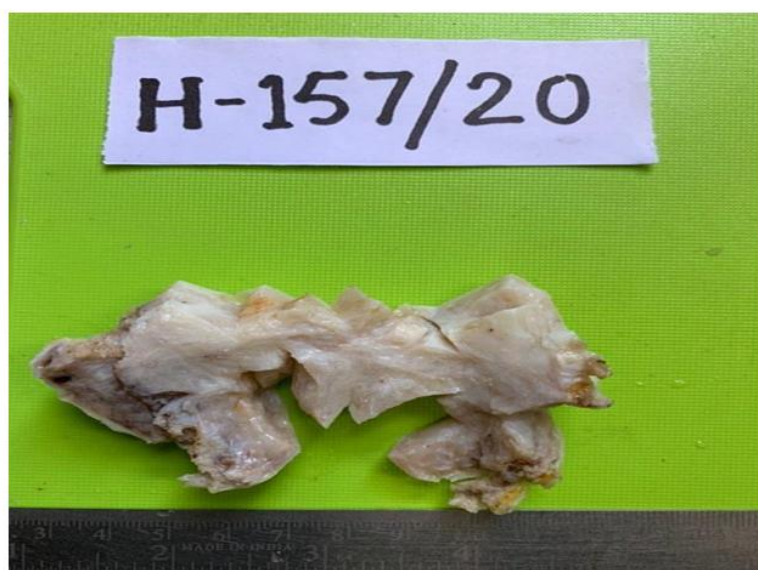


Figure 2: External surface of the tumour is smooth with greyish white to yellow solid and cystic cut surface.

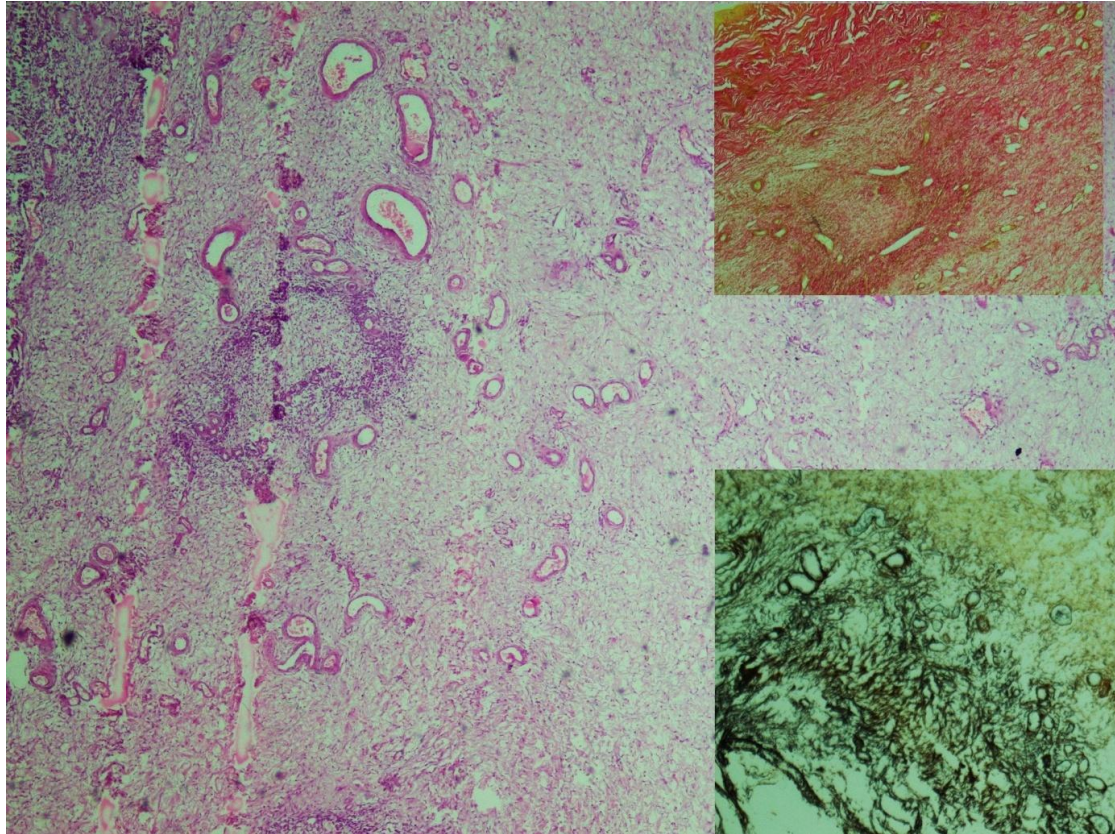


Figure 3: Microphotograph showing pseudo lobular appearance with prominent vascularity and edematous stroma. (H&E 10X) (inset highlighting collagen by VG and reticulin around individual tumour cell)

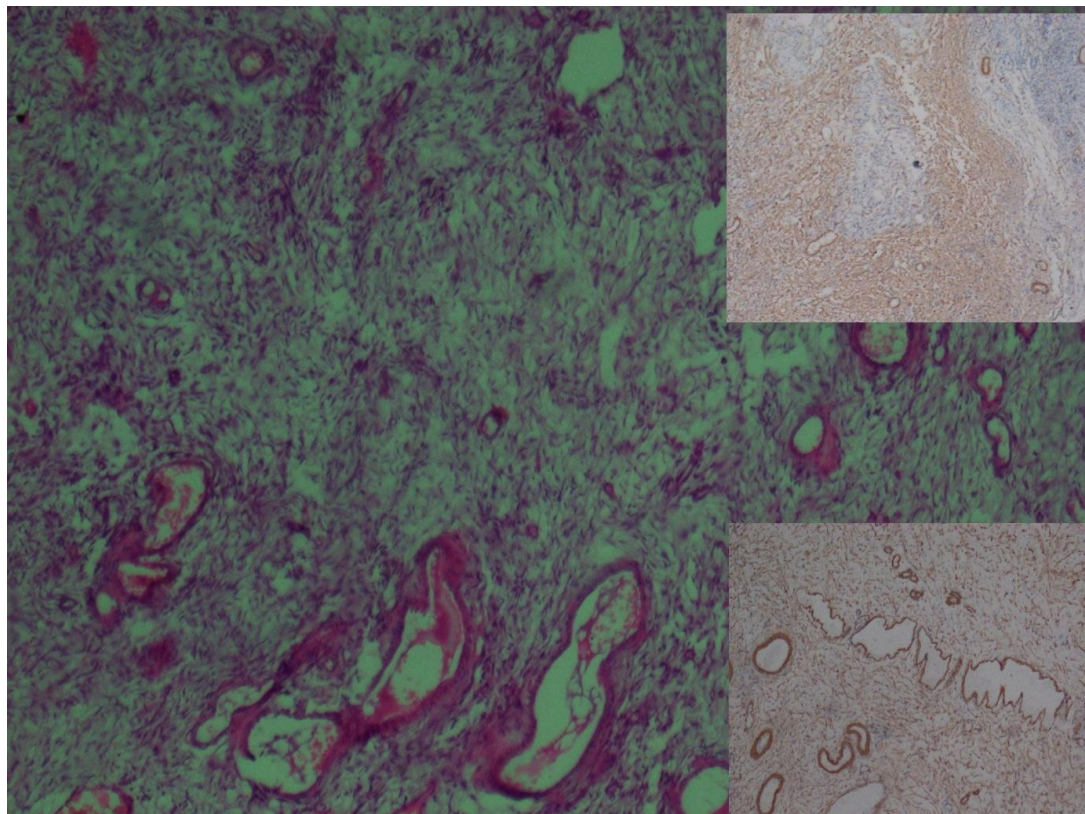


Figure 4: Microphotograph showing lobules with numerous blood vessels, many of them are ectatic. The stroma of the tumour is focally myxoid. (H&E 40X) (lower inset highlighting SMA and upper inset showing inhibin positivity around the vessels and Tumour cells)

Light microscopic examination revealed compressed normal ovarian stroma along with presence of a tumor with a pseudolobular architecture. There were seen cellular area composed of bland spindled cells with minimal atypia. Mitotic activity is inconspicuous. Cellular area are admixed with hypocellular area with collagenous, oedematous and myxoid stroma. There was prominent vascularity noted with in the tumor, with presence of dilated, thin walled vessels. There is no evidence of necrosis.(Figure 5)

The Van Gieson staining demonstrated abundant collagen in between the tumor cells. Reticulin staining demonstrated reticulin fibres around the individual cells. Immunohistochemically, the tumor was positive for smooth muscle actin (SMA) and inhibin.

Based on the above clinical and histopathological findings and IHC, a diagnosis of SST was established.

III. Discussion

Ovarian sex cord-stromal tumors are relatively infrequent neoplasms that account for approximately 8% of all primary ovarian neoplasms(5). Sex cord-stromal tumors of the ovary include granulosa cell tumors, fibrothecomas, Sertoli-Leydig cell tumors, steroid cell tumors, and SSTs. SSTs account for only 6% of sex cord-stromal tumors [2,4].

SST is distinctive from other sex cord stromal tumors because of its unusual clinical presentation at an early age and lack of hormonal manifestations. The most common clinical symptoms include menstrual irregularities, pelvic pain, and nonspecific symptoms related to an ovarian mass. Masculinization or anovulation may be present in some patients, as they are occasionally associated with estrogen and androgen secretion. SSTs usually present in the 2nd and 3rd decades of life; whereas, other ovarian stromal tumors present in the 5th or 6th decade of life[6,7]

The etiology of SSTs is not well understood. Ismail et al, suggested that an endocrine milieu might be responsible for the morphology of SSTs and that they may develop from pre-existing ovarian fibromas[8].

On the basis of ultrastructural features, SST were thought to be arise from the pluripotent immature stromal cells of ovarian cortex. However, now it is proposed that SSTs are derived from a population of muscle specific actin positive elements from the theca externa, namely, the perfollicular myoid stromal cells.[9]

Ultrasonography is a useful initial tool for differentiating between cystic and solid masses and determining the organ of origin. However, computed tomography and MRI are both more sensitive for delineating the nature of the mass and tumor extension. On MRI, a diagnosis of SST can be strongly suggested, when typical signal patterns such as hypointense nodules, hyperintense stroma, lobulation, strong enhancement with gadolinium, and a peripheral hypointense rim are present[10].

SST should be considered in the differential diagnosis of solid ovarian mass in adolescent, as it may affect planned surgical approach.

The differential diagnosis of SSTs of the ovary includes other sex cord-stromal tumors such as fibromas and thecoma, which generally occur in the fifth or sixth decades of life. These may be differentiated from the SST on the basis of histopathology and immunohistochemical findings[10]. SSTs are heterogenous characterized histopathologically by cellular pseudolobules, prominent interlobular fibrosis, marked vascularity, and a dual cell population: Collagen producing spindle cells and lipid-containing round or ovoid cells. The heterogeneity due to the variation in cellular size and shape are helpful features in the differential diagnosis of SST, and contrasts with the relative homogeneity of thecomas and fibromas. CD 34 stain the endothelium of the dilated and branched vascular architecture of SST and clearly distinguishes SST from thecoma and fibroma [5,11,12]. Immunohistochemical analysis in our patient demonstrated positivity for inhibin and SMA and negativity for desmin, S-100 protein, and cytokeratin.

Vascular tumors are included in the differential diagnosis due to prominent vascularity, which relate to elaboration of vascular endothelial growth factor (VEGF), but inhibin positivity suggests the diagnosis of SST.

Massive ovarian edema may be confused with SST. But preserved ovarian tissue within the edematous stroma and absence of heterogeneity favors the diagnosis of massive ovarian edema. In addition, the edema of SST is zonal in contrast to that seen in massive ovarian edema or an edematous fibroma.

Infrequently the vacuolated cells and the presence of signet-ring cells in association with edematous stroma may be mistaken for signet-ring cells of Krukenberg tumor of the ovary. But these malignant tumors occur typically in women in the sixth and seventh decades, are mostly bilateral and lack the pseudolobulated pattern of SSTs. Furthermore, signet-ring cells of Krukenberg tumors contain mucin rather than lipid, and they may exhibit mitotic activity and nuclear atypia[12].

SST can be treated successfully by enucleation or unilateral salpingo-oophorectomy. No local or distant recurrence noted in the literature [13].

IV. Conclusion

Due to the rarity of SSTs, it is not always possible to predict the presence of this tumor preoperatively on the basis of clinical and sonographic findings. It has a benign course and entails a very good prognosis with conservative surgical treatment. Characteristic histopathological features and IHC establish the diagnosis.

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