Case Report

Sclerosing stromal tumor of ovary in a young female: A case report and brief review of literature

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ABSTRACT

Sclerosing stromal tumor (SST) is a rare benign sex cord-stromal ovarian tumor typically encountered in young females. These tumors are hormonally inactive and usually present with nonspecific symptoms. Histopathology is essential in the diagnosis of this uncommon tumor. We present a case of SST in a young female with a review of current literature. This patient presented with abdominal pain. She underwent right salpingo-oophorectomy. The histopathological examination and immunohistochemistry of the tumor was consistent with the diagnosis of SST.

Key words: Ovarian tumor, ovary, sclerosing stromal tumor, sex cord-stromal tumor

INTRODUCTION

Sclerosing stromal tumor (SST) is an uncommon benign sex cord-stromal ovarian tumor. It represents 2–6% of all stromal tumors of the ovary. It is distinct from other sex cord-stromal tumor of the ovary because of its predominant occurrence in young women. It usually occurs below 30 years of age and lacks hormonal manifestations.[1,2] It presents mostly with menstrual irregularity and pelvic pain.[3] Histopathological confirmation is the cornerstone for the diagnosis of this uncommon tumor. Histologically, SST is characterized by cellular heterogeneity, prominent vascularization, and a pseudo-lobular appearance composed of alternating cellular and hypocellular areas.[2,4]

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To date, only a few cases of SST have been reported in the literature.[4-7]

We present this uncommon case of SST in a young female highlighting the importance of histopathological examination in the diagnosis of this benign ovarian tumor. We have also discussed the differential diagnoses along with a brief review of the literature.

CASE REPORT

A 16-year-old female presented with a complaint of abdominal pain for 1 month. The pain was described as having a squeezing character. The pain was localized to the right lumbar region. There was no history of vomiting, fever, loss of weight or loss of appetite. There was also no history of dysuria or difficulty in micturition. She was not a known patient of diabetes, hypertension, asthma or epilepsy. She had attained menarche at the age of 14 years.

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Her menstrual cycle was irregular. She did not report any history of previous surgery.

She had a soft palpable lump over the hypogastric region. The lump was mobile, nontender and was measuring 5 cm × 7 cm.

Computed tomography (CT) showed a right ovarian complex mass with solid and cystic component. The mass was heterogeneously enhancing, well defined and lobulated [Figure 1]. Routine hematological and biochemical investigations were within normal limits. Tumor markers like CA-125, alfa fetoprotein, and beta-human chorionic gonadotropin were within their normal limit.

She underwent right salpingo-oophorectomy. The external surface of the ovarian mass was smooth. The cut surface was solid and grey-white in appearance. The attached large uniloculated cyst contained serous fluid.

Histopathologically the cyst was a benign serous cyst. The solid area showed compressed normal ovarian tissue and a neoplasm comprising spindle cells and polygonal cells with minimal pleomorphism. The spindle cells were arranged in the form of fascicles with minimal nuclear atypia. Mitotic activity was inconspicuous. There was prominent vascularity noted within the tumor. Necrosis was not noted. There were areas of luteinized cells [Figure 2]. The Masson trichrome staining demonstrated abundant collagen in between the tumor cells [Figure 3]. Immunohistochemically the tumor was diffuse positive for vimentin. Smooth muscle actin showed positivity in the blood vessels, but was negative in the lesional component [Figure 4].

She had an uneventful postoperative period and is doing well after 1½ year of surgery.

DISCUSSION

SST is a rare benign ovarian neoplasm of stromal origin. It was described as a distinct entity by Chalvardjian and Scully in the year 1973.[5] The SSTs are unilateral tumors predominantly affecting females in the second and third decades.[3,7] Our patient was a 16-year-old female with right side unilateral SST. The status of the left ovary, as per the imageological findings was unremarkable.

The clinical symptoms of SST are nonspecific. The typical clinical presentation of SST is pelvic or abdominal pain accompanied by tenderness, a pelvic and/or abdominal mass, and/or menstrual irregularity. Usually, SSTs are hormonally inactive.[3,4,7] The most common presenting symptom in SST is a menstrual irregularity. SST patients on rare occasions have ascites.[8] Our patient presented with abdominal pain. She gave a history of the irregular menstrual cycle. She did not however have any hormonal symptoms and nor was there any ascites.

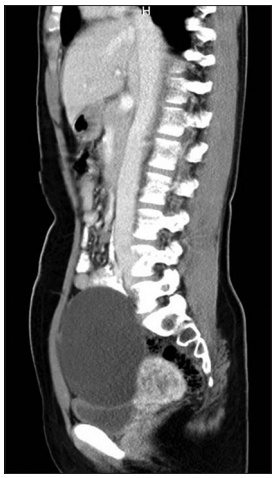

Figure 1: Contrast-enhanced sagittal reconstruction images of right solid and cystic ovarian lesion

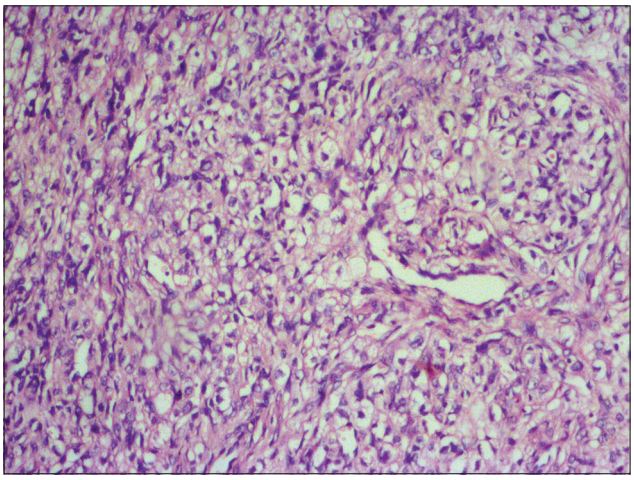

Figure 2: Tumor showing polygonal cells with prominent vascularity (H and E, ×200)

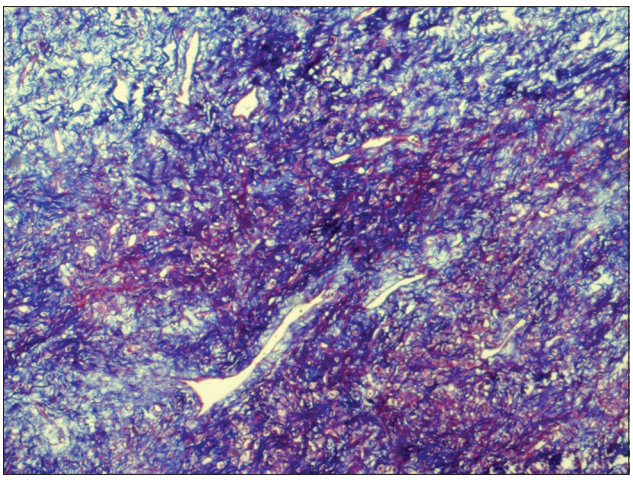

Figure 3: Cellular areas of tumor showing collagen (Masson trichrome, ×40)

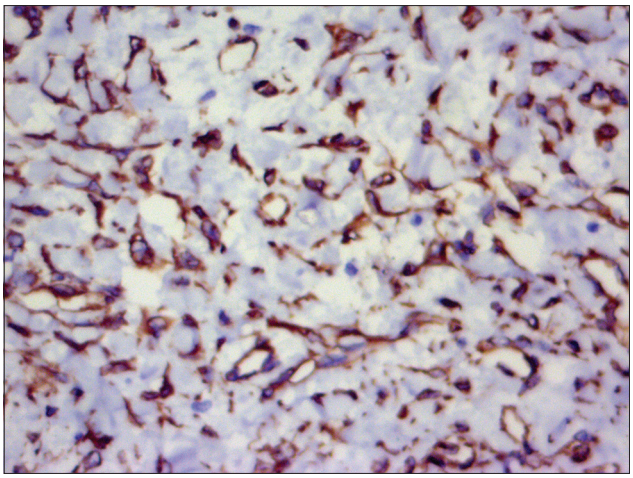

Figure 4: Immunohistochemistry showing positivity for vimentin (IHC, ×200)

Imaging studies are useful. They frequently reveal solid or complex cystic adnexal masses with marked vascularity.[9] On CT scan, our patient showed right ovarian mass lesion with cystic and enhancing solid component.

The gross appearance of SST ranges from small solid masses to large multicystic masses.[4] The histopathological examination helps in the definitive diagnosis of SST.[3,4] Microscopically, these tumors give a pseudo-lobular appearance of alternating cellular and hypocellular areas. The cellular areas comprise of spindle cells with dense collagenous tissue. In the hypocellular areas, there are clusters of lutein cells characterized by round to ovoid cells with perinuclear vacuolation. Also seen are areas of sclerosis and prominence of blood vessels.[3,4,7]

In our case, the compressed normal ovarian stroma was present peripherally. The lesion was circumscribed and showed cellular spindle cell areas and polygonal vacuolated cells. The lesion had rich vascularity with the presence of branched vessels. Masson trichrome stain demonstrated abundant collagen in between tumor cells in the cellular area.

Immunohistochemical staining is usually not required for the definitive diagnosis of SST. All SSTs typically show positive staining for inhibin and calretinin, alfa-glutathione S-transferase (αGST) and positive CD34 staining of the endothelium of the branching blood vessels.[7]

The important differential diagnoses include other sex cord-stromal tumors like thecoma, fibroma and lipoid cell tumors. Sometimes vascular tumors, massive ovarian edema, and rarely Krukenberg's tumor can be considered in the differential diagnoses. SSTs are heterogeneous whereas other stromal tumors are homogenous. SSTs and thecomas and fibromas show positivity for inhibin. But, CD34 stains the endothelium of the dilated and branching vascular architecture of SST and clearly distinguishes SST from thecoma and fibroma. αGST shows positivity within scattered cells in SST, diffuse staining in thecomas and no staining in fibromas. The SSTs are positive for inhibin in contrast to tumors of vascular origin.[7] Sometimes benign condition like massive ovarian edema can be considered in the differential diagnosis but it lacks the heterogeneity of SST and shows preserved ovarian tissue within the edematous stroma.[7] Sometimes lutein cells of SSTs exhibit signet ring-like cells mimicking that of Krukenberg's tumor. The signet ring cells of Krukenberg's tumor contain mucin rather than lipid, and the cells may exhibit nuclear atypia and mitotic activity. Krukenberg's tumors are mostly bilateral, occur usually in the sixth and seventh decades and lack pseudo-lobular pattern of SST.[4,7,10]

In our case, vimentin showed diffuse positivity thereby indicating stromal origin. Other immunomarkers like smooth muscle actin and CD34 showed positivity only in the blood vessel walls. Inhibin was not done in our case. However, the typical histopathological findings with added demonstration of collagen by special stain and positivity for vimentin and CD34 helped in the diagnosis of SST.

It is difficult to diagnose SST by only by clinical and radiological examination as these tumors are rare. But in a young patient presenting with an ovarian mass, SST should be considered as one of the diagnoses. Intra-operative frozen sections can help to diagnose SST by its pseudo-lobular appearance, heterogeneous cellular areas, and stromal hyalinization or edema. SST is a benign tumor and can be treated successfully with fertility preserving surgery.[4]

CONCLUSION

This uncommon case of SST of the ovary in a young female emphasizes the importance of histopathological examination in the diagnosis of this benign ovarian tumor. This is an additional case report of SST occurring in a young girl.

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Conflicts of interest

There are no conflicts of interest.

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