

Bilateral Sclerosing Stromal Ovarian Tumor: A Rare Case Report

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Abstract

Sclerosing stromal tumor of the ovary is believed to be a rare, benign, sex cord stromal tumor occurring predominantly in younger women in the 2nd and 3rd decades of their life. The prevalent clinical presentations include pelvic or abdominal pain, mass, or menstrual changes. Even though it is occasionally accompanied by hormonal manifestations, virilization as a result of androgen production by the tumor is rare. The present research was conducted to present a rare case of bilateral sclerosing stromal ovarian tumor in a 20-year-old patient with irregular menstruation and pelvic pain. All of the sclerosing stromal tumors were benign and were treated successfully with enucleation or unilateral oophorectomy.

Keywords: Ovarian tumor, Sclerosing stromal tumors, Case report

Introduction

Sclerosing stromal tumor (SST) of the ovary is a rare benign classification as a sex cord stromal tumor.¹ This was primarily described by Chalvardjian in 1973.² The majority of cases occur in female patients under the age of 30 at the mean age of 28 years.³ They are typically unilateral and well-circumscribed.⁴ Bilateral presentation is rare and reported in only four cases.⁵⁻⁸ Naidu et al. reported a 14-year-old girl, who had referred for evaluation of primary amenorrhea and the absence of breast

development. Physical examination revealed a lean, non-dysmorphic female without acne or hirsutism. The abdominal examination was normal.⁵ Additionally, Gilani et al. reported a 19-year-old female presented with two months history of pelvic pain. The computed tomography (CT) scan of abdomen and pelvis presented a septated, multicystic mass apparently arising from adnexal region inferiorly, and deforming deviating uterus.⁶ Chang et al. studied an 11-year-old premenarchal girl. The physical

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examination revealed a large, solid, immobile, non-tender mass in the suprapubic region. The patient was Tanner stage III without evidence of virilization.⁷ Moreover, another case report introduced a 12-year-old premenarchal girl presented with a palpable mass in the lower abdomen. The physical examination revealed the presence of a large, firm palpable mass with distension in the mid- and lower abdomen. The girl was in Tanner stage III and was premenarchal.⁸

SST is normally presented with pelvic or abdominal pain and tenderness, a mass, and/or abnormal menses.⁹ In certain cases, it is hormonally active and can cause virilization, hirsutism and rarely endometrial carcinoma.⁵ Diagnosis of SST is often made by post operative pathologic examination. The important differential diagnosis are other sex cord stromal tumors, including fibroma, thecoma, and lipoid cell tumors.^{10,11}

Herein, we presented the fifth reported case of bilateral SST in a 20-year-old female and described our findings concerning ultrasound (US) and magnetic resonance imaging (MRI).

Case Presentation

A 20-year-old girl referred with irregular menses and abdominal pain from two years ago.

Bilateral adnexal mass was found in the pelvic examination. Tumor markers, including carcino embryonic antigen (CEA), alpha fetoprotein (AFP), beta-hcG, LDH, were within normal limits. Based on the clinical examination, she was moderately anemic.

Abdominopelvic sonography revealed mixed echogenic mass measuring 10³×51 mm, including fat and calcification foci in her right ovary and 33×17 mm solid echogenic nodule in her left ovary.

A followed gadolinium enhanced MRI of pelvis revealed the presence of two round mass lesion measuring 56 mm and 42 mm in the right pelvic side, which ere isosignal on T1 imaging and low signal intensity on T2 imaging, including high signal foci. The post-contrast images demonstrated enhancement, which indicated pedunculated myoma. Moreover, 20 mm mass lesion was seen in the left ovary, which was low signal on T1 and T2 imaging. The post contrast images showed faint enhancement (Figures 1 and 2). An accurate uterus was identified.

Intraoperative findings at laparotomy revealed abnormal appearance of both ovaries including multiple hard, irregular border nodulous mass measuring 2×2 and 3×3 cm in the left ovary, which were resected. There was a mass of 10 cm

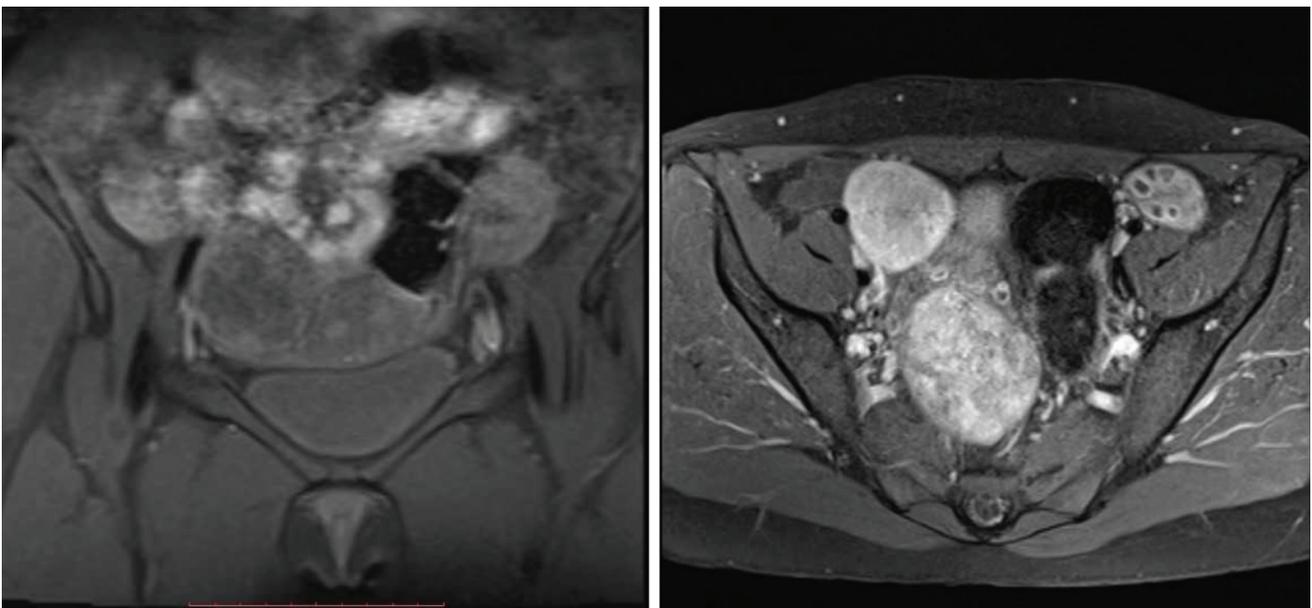


Figure 1. (a) Axial: T1 weighted imaging shows a non-homogenous iso intense mass in the right adnexa, (b) the left adnexal mass with vivid enhancement on post contrast axial T1 weighted images.

diameter originating from the right ovary, which was also resected. In addition, a 7×6 cm lobulated uterine tumor that was attached to the right ovary was enucleated. No other tumors were observed in the peritoneal cavity. Both ovaries and uterus could be salvaged.

Histopathology report

Gross examination

The specimen consisted of about eight nodular pieces of tissue measuring about 2×2.5 cm from both the ovaries. The cut surface showed yellowish color and elastic area with hard foci.

Gross examination of 7×6×5 cm uterine mass revealed an elastic tumor with lobulated outer surface with yellowish area on the cut surface.

Microscopic study

Ovarian nodules exhibited spindle cell proliferation with fascicular pattern. Some foci showed collagen fiber without cytonuclear atypia, associated with round to ovule cells with acidophilic to clear cytoplasm. Vascular space was also observed with some irregular features.

Immunohistochemical (IHC) analysis on tissue blocks (ovarian pieces) revealed positivity for actin and weakly positive for ER, while negativity for S 100, WT1, CD 34, EMA, desmin and inhibin was also represented.

IHC finding were suggestive for SST of ovary (Figure 3).

Discussion

Sex cord stromal tumors represent approximately 8% of ovarian neoplasm.¹² SST

compromises less than 5% of sex cord stromal tumor and appears to occur frequently in the second or third decades of a woman's life. The patients suffering from this kind of tumor may have irregular menstruation, menorrhagia, or lower abdominal pain.¹³ Final diagnosis of SST is usually confirmed following pathologic examination after surgery. SSTs usually have portion with higher cellularity, stromal portion dominantly with collagen, and edematous portion, so that pathologists could distinguish it from fibroma, thecoma, or lipid cell tumors.¹⁴

MRI is helpful in differentiating SST from malignant ovarian tumors, which include a mass with hyper intense cystic components or a heterogenous solid mass of intermediated to high signal intensity centrally and intermediate to low signal intensity peripherally on T2 weighted MRI. Dynamic contrast - enhanced T1 images are even capable of distinguishing SST from other sex cord stromal tumors with striking early peripheral enhancement (intermediate signal) reflecting cellular areas with prominent vascular networks and an area prolonged enhancement in inner portion of the mass (low signal) representing collagenous hypocellular area.

This illustrates that MRI is useful in making a preoperative diagnosis of SST and distinguishing SST from other malignant ovarian tumors, as well as other stromal tumors.¹⁵

The peripheral like band, for instance low signal intensity on T2 weight images may reflect compressed ovarian tissue, which is a



Figure 2. (a) Axial: T2 weighted image represents the left adnexal hypointense mass with homogenous texture, (b) T2 TIRM MR image in sagittal plane shows a non-homogenous mass with high signal and iso signal area, (c) The left ovary mass with round shape and low signal intensity on T2 weighted images.

distinguishing feature between SST and fibroma/thecoma. The high signal intensity area on T2 weighted images correlate with poorly cellular tissue (fibrous portion of the tumor). Early peripheral enhancement with centripetal progression and general lack of enhancement within the central area are usually seen on post contrast images.¹²

The unusual fact about our patient was the T1 and T2 images of her left ovarian mass showing

decreased signal abnormality within the central portion of the mass whereas post contrast images, showed faint enhancement that indicated fibrous portion of tumor. In addition, the two right masses were hyposignal with hyper signal foci on T2 images; post-contrast images indicated homogenous texture in both masses.

Histologically, SST is characterized by cellular heterogeneity, prominent vasculature and a pseudolobular appearance composed of both

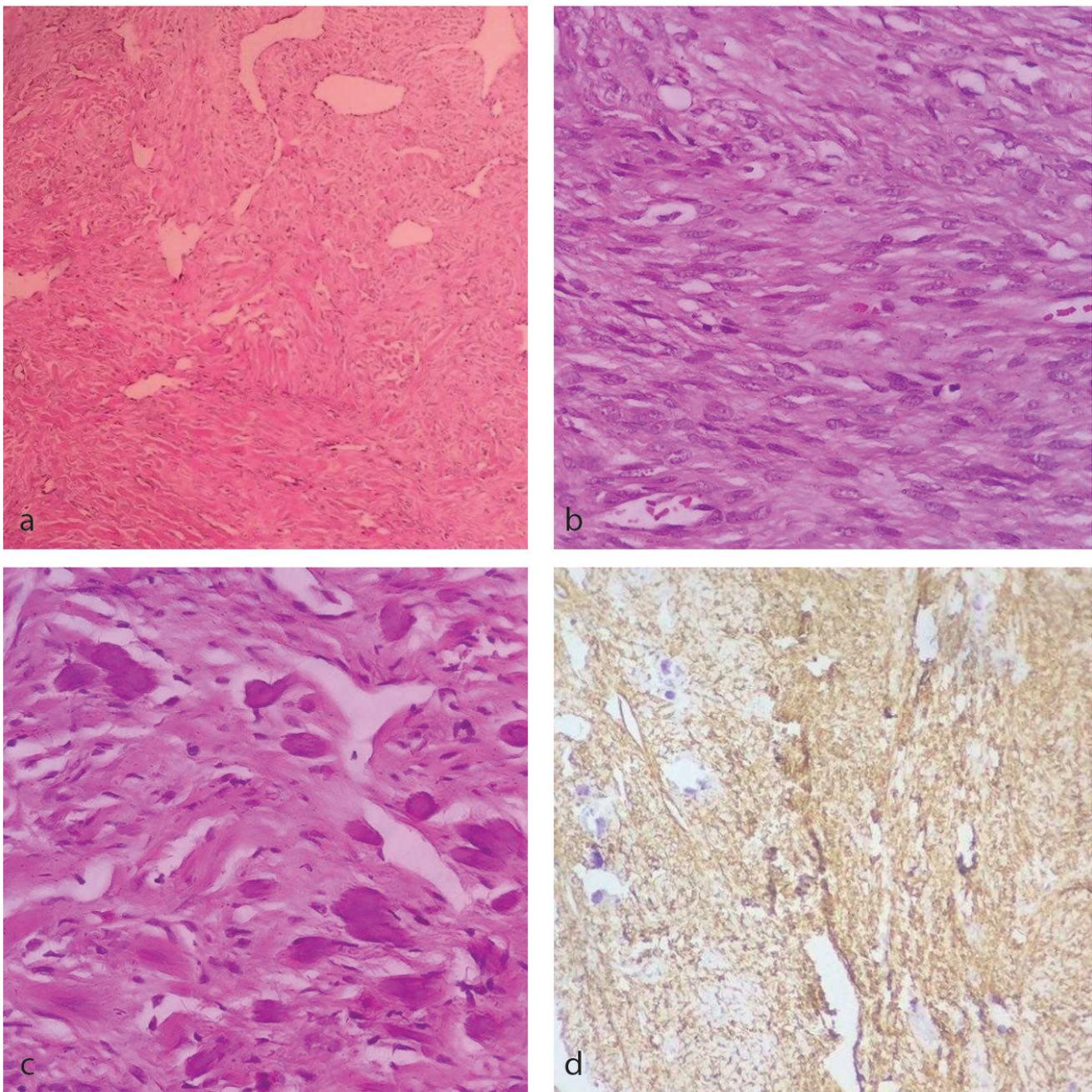


Figure 3. (A&B) cellular areas composed of collagen producing spindle cells and round / oval cells with marked vascularity (H&E, $\times 100$), C) Area of calcification(H&E, $\times 100$), D) immunohistochemistry staining , diffuse staining for actin.

cellular and hypocellular areas. Positive vimentin reaction, weakly positive desmin and smooth muscle, specific positive actin stains and a negative cytokeratin stain in SST have been reported.¹⁶ IHC of desmin and smooth muscle actin is useful for distinguishing SST from the fibroma.¹⁷ We emphasize the significance of being familiar with sclerosing stroma tumors when evaluating ovarian neoplasm in children and adolescents, in order to reach an appropriate clinical management preventing extensive and unnecessary surgery and preserving fertility.

Conclusion

Due to the rarity of this ovarian neoplasm, it is not always possible to predict the presence of this tumor preoperatively, yet young patients with ovarian mass need to be educated about this rare tumor. Being well-informed helps to prevent extensive morbid surgery, as all of the sclerosing stromal tumors were benign and were treated successfully with enucleation or unilateral oophorectomy.

Informed Consent

We obtained written informed consent from the patient for the publication of this case report and any accompanying images.

Conflict of Interest

None declared.

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