Sclerosing Stromal Tumor of Ovary

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Sclerosing stromal tumor of the ovary is a rare ovarian disease with prevalence of 1.5% to 6% of ovarian stromal tumors. We present a 24-year-old woman with irregular menstruation for 6 months and a self-palpable lower abdominal mass. Enucleation of the left ovarian tumor was undertaken. Gross examination showed a soft elastic tumor with a smooth outer surface and diffusely white edematous stroma with scattered yellowish nodular areas on the cut surface. Histologic study showed that the cellular nodules consisted of a disorganized admixture of fibroblasts and round vacuolated tumor cells. The cellular nodules were separated by edematous hypocellular fibrous tissue forming a pseudolobular appearance. Regular menstruation resumed after the surgery and no evidence of tumor relapse during 1 year of follow up. *(Chang Gung Med J 2003;26:444-8)*

Key words: Sclerosing stromal tumor (SST), ovarian neoplasms, sex cord-stromal tumor.

Sclerosing stromal tumor (SST) was first delineated as a distinct entity in the ovarian sex cord stromal tumors in 1973 by Chalvardjian and Scully.[1] SST is a benign neoplasm and is distinguished from other ovarian stromal tumors by the production of collagen and a pseudolobular pattern with cellular areas separated by edematous and collagenous areas.

CASE REPORT

A 24-year-old woman, gravida 2, para 0, had abdominal fullness and a self-palpable low abdominal mass for 1 month. She had irregular and frequent menstruation every 15 to 20 days for 6 months. Physical examination revealed a solid, non-tender tumor over the left adnexal area, and pelvic ultrasounography with color doppler showed a well-defined solid mass measuring 8.4×7.0 cm over the left adnexa and a small amount of ascites (Fig. 1). The tumor contained hyperechoic honey-comb structures with a resistance index (RI) of 0.47 and pulsatility index (PI) of 0.68 over the tumor artery. Her serum CA125 level was 194.22 U/ml (reference range, <35 U/ml). Serum carcinoembronic antigen, alpha-fetoprotein, and beta-human chorionic gonadotropin were within reference ranges. Hormone assay including serum follicle-stimulating hormone (FSH), luteinizing hormone (LH), thyroid-stimulating hormone (TSH), prolactin, estradiol,

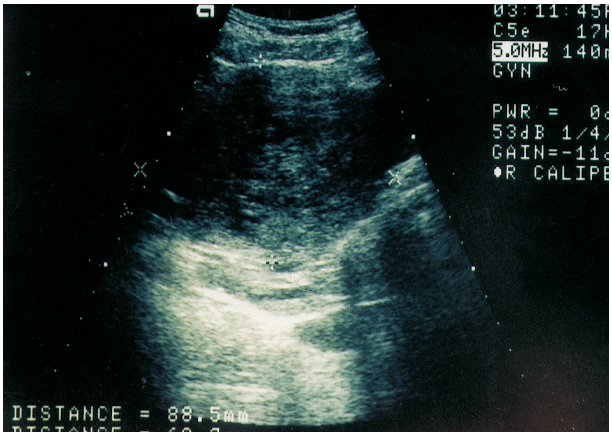

Fig. 1 A well-defined solid ovarian mass on the left side, measuring 8.4×7.0 cm in dimension.

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progesterone and testosterone were also within reference ranges. She had no gastrointestinal symptoms and no gastric tumors were seen under panendoscopy.

Under the impression of a solid ovarian tumor, the patient underwent laparotomy. After opening the peritoneal cavity, 30 ml of clear fluid in the cul-de-sac was found. A smooth, well-circumscribed, bosselated left ovarian tumor of 11.8×8.6 cm in dimensions was enucleated. The uterus, the right ovary and bilateral fallopian tubes were normal in appearance. There were no other tumors in the peritoneal cavity. Gross examination showed a soft elastic tumor with smooth outer surface and diffusely white edematous stroma with scattered yellowish nodular areas on the cut surface (Fig. 2). Microscopically, it showed a pseudolobular appearance composed of cellular nodules that were separated by edematous hypocellular fibrous tissue (Fig. 3). The cellular nodules consisted of a disorganized admixture of fibroblasts and round vacuolated cells. Prominent vascularity were present in these areas (Fig. 4).

After an uneventful post-operative recovery, regular menstruation resumed and has continued until the writing of this manuscript, 24 months after treatment. Regular follow up showed no evidence of tumor relapse.

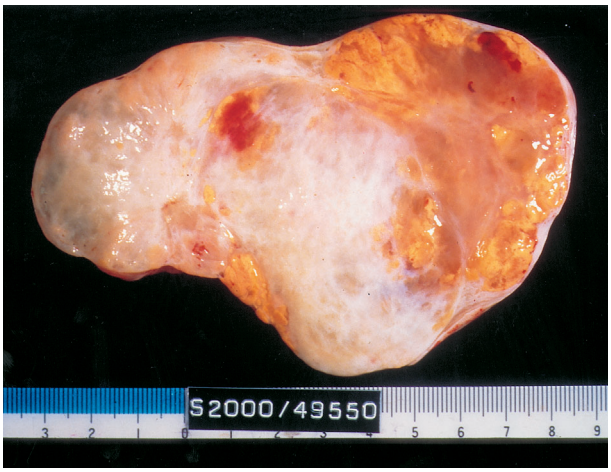

Fig. 2 Cut surface of the left ovarian tumor show a diffusely white and edematous stroma with scattered yellowish nodular areas in the peripheral.

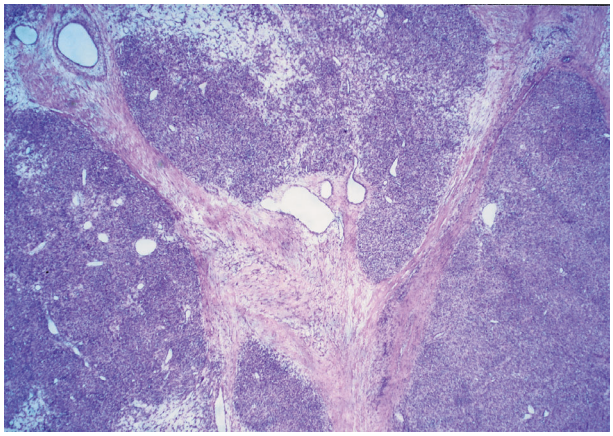

Fig. 3 Low power field show a pseudolobular appearance composed of cellular nodules separated by edematous hypocellular fibrous tissue. (H&E stain, 40×)

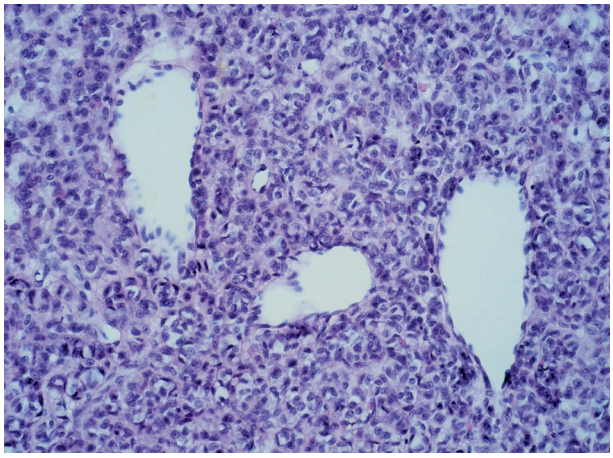

Fig. 4 High power field of the cellular nodules show admixture of fibroblasts and round vacuolated cells with some resembles signet-ring cells. Prominent small capillaries are present in these cellular nodules. (H&E stain, 150×)

DISCUSSION

SST of the ovary is a distinct subtype of the ovarian stromal tumors. We undertook a MEDLINE search using keywords ovarian neoplasms and sclerosing stromal tumor to obtain reports on this tumor in the English literature and then extended the search to related reports listed in their references. We concluded that up to the writing of this paper, a total of 114 cases had been reported.[2-12] All cases were diag-

nosed as benign except for one patient with low-grade malignancy reported in 1990.[13] The patient presented with sudden onset of abdominal pain and histologic examination showed a mitotic index of 4 mitoses per 10 high-power fields (HPF), and a SST with low-grade malignancy was diagnosed. Further follow up information was not available in the report.

Ovarian SST occurs more commonly during the second to third decades of life with an average age of occurrence of 27.5 years. More than 80% of SSTs occur in patients below the age of 30 years.[14] The most common presenting symptoms are menstrual irregularities and pelvic pain. Palpable lower abdominal tumors and hirsutism have also been reported.[3,4] Tumor size varies from 1.5 cm to 20 cm in diameter.[5] Elevated serum CA125 level and/or ascites were depicted in some cases.[2,3,6] The serum levels of 17-β-estradiol, progesterone and testosterone were within reference ranges in our patient and these findings are consistent with the finding reported by others.[6]

Although many patients presented with menstrual abnormalities, only a few had documented biochemical evidence of hormone production of the tumor. In addition, both elevated estrogenic and androgenic hormone production were described by Damajanov et al.[15] Infertility and endometrial hyperplasia concomittent with SST have also been described which might indicate a status of excessive hormone production.[16] Other authors have documented elevated levels of both estrogenic and androgenic hormones that were corrected after surgery. In several patients with irregular menses,[7,8,12,13,15] normal menses following the excision of the tumor was noticed. In our patient, the hormone levels were normal and the patient had no clinical virilization. Her menstruation returned to normal after surgery.

Sonographic findings of SST showed a well-defined solid mass with hyperechoic honeycomb structures in our patient, which are also the characteristics of mixed heterogenecity tumor without focal calcifications.[17] In our patient, the resistance index of the tumor artery was 0.47 and the pulsatility index was 0.68. A case presented in another report showed the resistance index was 0.50 and the pulsatility index was 0.71.[18]

Preoperative diagnosis may be possible based on magnetic resonance imaging (MRI) findings that demonstrated the characteristics of SST included pseudolobulation, which consists of low-intensity nodules set against high-intensity stroma on T2-weighted images.[2] The presence of tightly packed cellular areas associated with foci of sclerosis explains the low density of these nodules on T2-weighted images. High-intensity areas on T2-weighted images correlated with poorly cellular tissue that was markedly edematous. However, the distinction of other stromal tumors and metastatic ovarian tumors to SST on MRI need further investigation.

Histologically, SST is characterized by cellular heterogeneity, prominent vasculature, and a pseudolobular appearance composed of both cellular and hypocellular areas.[3] The descriptive term of "sclerosing stromal tumor" was proposed because the cellular areas of this tumor tend to undergo collagenous sclerosis. It has occasionally been confused with massive ovarian edema and Krukenberg's tumor. The distinction between SST and Krukenberg's tumor depends on immunohistochemistry stain.

Twelve cases of SST were analyzed immunohistochemically and demonstrated expression of vascular permeability factor/vascular endothelial growth factor (VPF/VEGF) in the luteinized theca-like cells and the receptor in capillaries and even in medium sized blood vessels.[3] Reverse transcription-polymerase chain reaction also showed the expression of VPF/VEGF messenger ribonucleatic acid in these tumors. Accordingly, the characteristic vasculature and edema of SSTs were considered to be associated with the expression of VPF/VEGF. In addition, three copies of chromosome 12 in 13-21% of all examined SSTs tumor cells was reported using fluorescence in situ hybridization (FISH) analysis.[3] One researcher described a patient with SST with monosomy of chromosome 16.[13] In addition, positive vimentin reaction, weakly positive desmin and smooth muscle-specific actin stains, and a negative cytokeratin stain in SST have been reported.[4] Immunohistochemistry of desmin and smooth muscle actin is useful in distinguishing SST from thecofibroma.[5]

In conclusion, though SST is a rare tumor, it should be considered in young woman with menstrual irregularity and pelvic mass. Enucleation is enough for this tumor and menstrual regularity resumes after surgery in most cases. No recurrence has been reported in our patient until now.

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卵巢硬化性間質瘤

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卵巢硬化性間質瘤是一種罕見的良性卵巢瘤，佔卵巢間質瘤的1.5-6%。我們報告一位24歲女性病人，因爲月經不規則及腹脹，發現此一腫瘤。病人接受腫瘤摘除手術，此腫瘤外觀平滑而且柔軟，縱切面可見白色水腫之間質，其間有散佈之黃色結節。病理組織切片顯示細胞結節被水腫之纖維組織所分隔，爲一卵巢硬化性間質瘤。手術之後，病人月經回復正常，而且經過一年的追蹤並無腫瘤復發之情形。(長庚醫誌 2003;26:444-8)

關鍵字：硬化性間質瘤，卵巢腫瘤，性索間質瘤。

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