Chulalongkorn Medical Journal

Volume 43
Issue 3 *March 1999*

Article 5

3-1-1999

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Sclerosing stromal tumor of the ovary: A case report

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Trivijitsilp P, Trirattanachart S, Sirisabya N, Niruthisard S. Sclerosing stromal tumor of the ovary: A case report. Chula Med J 1999 Mar; 43(3): 169-75

Sclerosing stromal tomor (SST) is an uncommon ovarian neoplasm occurring predominantly in young women. This case report describes a 20 year old patient who presented with irregular menstruation for the prior 3-4 years. Pelvic examination and ultrasonography revealed a left solid ovarian tumor . A left salpingoophorectomy was performed. A distinctive microscopic histology characterized by pseudolobulation of the edematous or collagenous hypocellular areas and the cellular areas with prominent vasculature of hemangiopericytomatous pattern distinguished this tumor from fibroma, thecoma and others forms of ovarian stromal tumor. Some investigations about hormonal function, immunohistochemistry and cytogenetic analysis have been reported.

Key words: *Sclerosing stromal tumor, Ovary.*

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Received for publication. January 20, 1999.

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Sclerosing stromal tumor (SST) เป็นเนื้องอกรังไข่ที่พบได้น้อยมาก โดยมักจะเกิดในสตรีวัย 11-30 ปี ผู้ป่วยหญิงรายนี้อายุ 20 ปี มาโรงพยาบาลด้วยอาการระดูมาไม่สม่ำเสมอ และกะปริดกะปรอย ตรวจภายในและคลื่นเสียงความถี่สูงพบเนื้องอกรังไข่ข้างซ้าย ได้ทำการผ่าตัดปีกมดลูกและรังไข่ข้างซ้ายออก ผลการตรวจทางพยาธิวิทยาพบลักษณะเฉพาะของเนื้องอกรังไข่ชนิดนี้ คือ ลักษณะ Pseudolobulation ที่ประกอบด้วย 1. บริเวณเนื้อเยื่อที่มีเซลล์หนาแน่นร่วมกับหลอดเลือดที่เรียงตัวคล้าย-รูป hemangiopericytomatous มากมาย 2. บริเวณเนื้อเยื่อที่มีเซลล์น้อย และบวมหรือประกอบด้วยคอลลาเจน ซึ่งช่วยให้วินิจฉัยแยกโรคนี้ จาก fibroma, thecoma และ ovarian stromal tumor อื่น ๆ ได้ มีการศึกษาการสร้างฮอร์โมน, immunohistochemistry, และความผิดปกติของโครโมโซมในเนื้องอกชนิดนี้ ซึ่งจะกล่าวต่อไป

In 1973, Chalvardjian and Scully first used the term " sclerosing stromal tumor " (SST) for an ovarian neoplasm which had a highly characteristic feature of collagenous sclerosing tendency in cellular areas of this tumor.[1] These distinctive histologic features distinguish SSTs from other ovarian stromal tumors (fibroma, thecoma, lipid cell tumor) and Krugkenberg tumor. It is a rare, benign ovarian tumor with a relatively small number of case reports.[1-12] Some further investigations for endocrine function, immunohistochemistry, and cytogenetic study had been reported[2, 4-9, 11] to identify histogenesis and morphogenesis of this tumor which will be discussed.

Case report

An unmarried 20 year old Thai female presented in December 1997 complaining of menstrual irregularity (metrorrhagia) for 3-4 years. She had been treated with hormonal drugs or contraceptive pills off and on by several general practitioners. Pelvic examination revealed a solid firm left ovarian mass about 7 cm in greatest diameter which was confirmed by transabdominal ultrasonography. Other physical examinations, laboratory investigations and chest X-rays revealed normal findings. At laparotomy, a solid firm left ovarian tumor measuring 8 x 5 cms was found and a left salpingo - oophorectomy was performed. The contralateral ovary, both tubes and uterus were quite normal.

Pathological findings

Grossly, the ovarian mass, measuring 8 x 5 x 5 cm^3, was oval, well circumscribed, and with a lobulated smooth surface and firm consistency. The cut surface was sharply demarcated, grey yellow lobulation intermingled with grey - white firm or edematous areas (Figure 1).

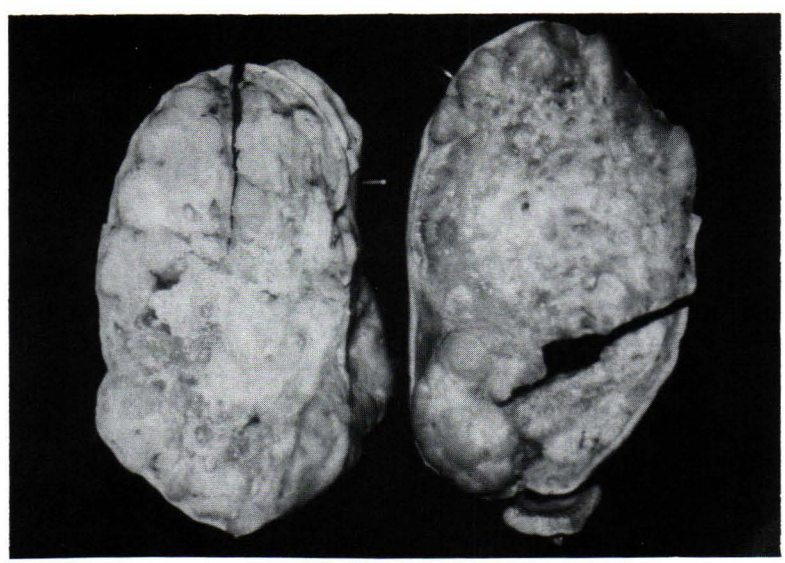

Figure 1. Gross section; showed well circumscribed mass with grey yellow lobulation.

Under light microscopy the sections revealed demarcated, pseudolobulated and hypercellular tumors surrounded by collagenous or edematous hypocellular stromal areas (Figure 2). The hypercellular areas exhibited a heterologous cell population consisting of luteinized thecalike cells with clear cytoplasm and round nuclei, as well as spindle - shaped fibroblastlike cells with eosinophilic cytoplasm and elongated nuclei. The small to medium - sized and thin walled dilated blood vessels in the cellular area were very prominent and showed a hemangioperi - cytomatous "staghom" configuration (Figure 3). Occasionally, some tumor cells in cellular areas displayed signet - ring like cells containing clear cytoplasm and compressed peripheral neclei (Figure 4) but were negative for periodic acid - Schiff (PAS) and mucicarmine stains. Immunohisto-chemical studies showed negative for estrogen and progesteronere ceptors.

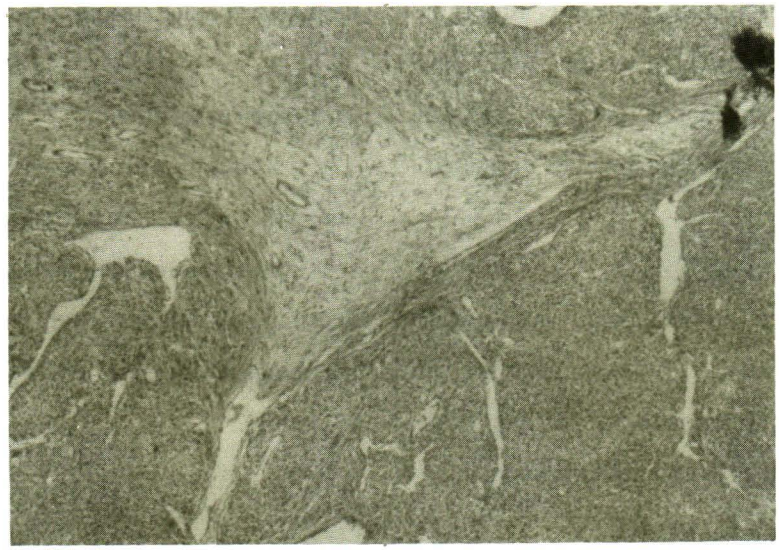

Figure 2. Edematous hypocellular stroma surrounds demarcated cellular areas associated with thin walled vessels creating the impression of pseudolobules.

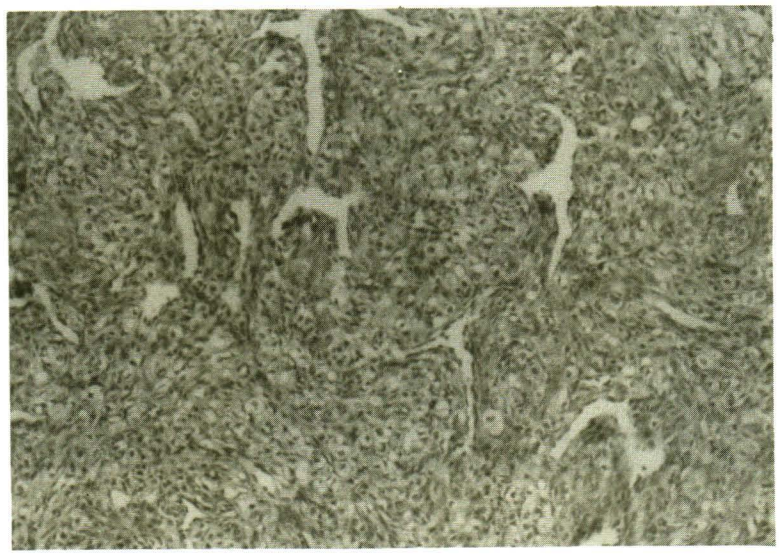

Figure 3. Prominent thin walled dilated vessels is seen in cellular areas which compose of spindle - shaped cells and rounded cells with clear cytoplasm.

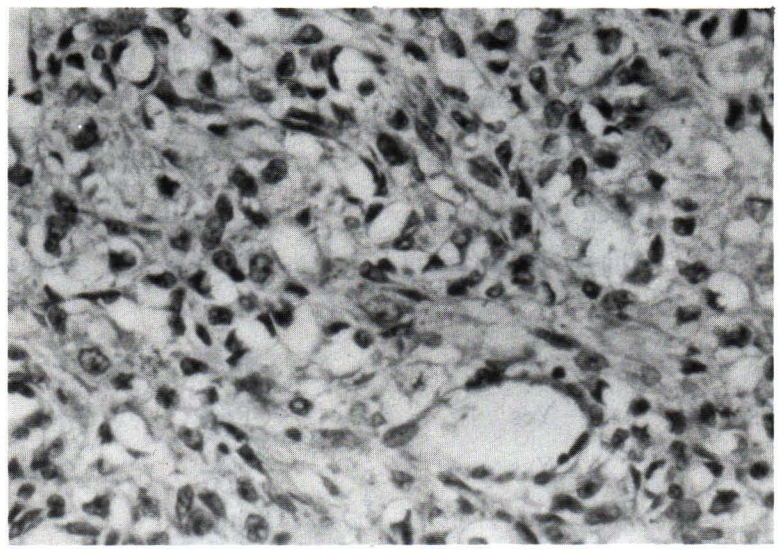

Figure 4. The compressed peripheral nuclei and clear cytoplasms result in signet - ring like cells which should not be confused with Krugkenberg tumors.

Discussion

Sclerosing stromal tumor of the ovary is classified as an uncommon neoplasm sex cord stromal tumor[1,2] with a relatively infrequent incidence of 4.1% of all ovarian neoplasms.[13] Therefore, it is a very uncommon neoplasm, having not more than 100 cases reported.[1-12] Eighty percent of cases occurred in patients below 30 years old with a surprising lack of hormonally related abnormalities,[1,2,14-16] but most cases have symptoms of menstrual irregularity[1,14,15] as in our case. However , some reports showed estrogen or progesterone or testosterone function.[7,12,17,18]

The distinctive microscopic features are characterized by a pseudolobular pattern of cellular zones separated by broad swathes of acellular collagenous or edematous connective tissue.[1,14-16] The cellular areas are comprised of polyhedral clear cytoplasmic cells and spindle-shaped cells intermingled with a hemangioperi - cytomatous pattern of thin walled dilated blood vessels.[1,14-16] Immunohistochemical studies revealed marked positive to smooth muscle actin, desmin, and vimentin [8,9,12,15] which suggests that this tumor should arise from perifollicular myoid stromal cells (a population of muscle-specific actin-positive cells in the theca externa).

Lopes, et al, showed monosomy 16 by cytogonetic studies[5] but Kwauchi, et al, revealed trisomy 12 in 13 - 21% of all examined SST cells and the association of the characteristic vasculature with the expression of vascular permeability factor and vascular endothelial growth factor.[12]

Sclerosing stromal tumor must be differentiated from other stromal cell tumors of the ovary, especially fibroma and thecoma.[1,14-16] the comparison of these tumors is summarized in Table 1 using the distinctive clinical and histological features of SST[14,16] which do not present in fibroma or thecoma . SST will not contain hyaline plaques which are common in the other ones. The other tumor that might cause difficulty in differential diagnosis is the Krugkenberg

Table 1. Sclerosing stromal tumor versus other stromal cell tumors.[14,16]

	Sclerosing stromal tumor	Fibroma	Thecoma
Age	80 % under 30 year (mean age 25 years)	90 % over 30 year (mean age 48 years)	
Function	Almost always absent	Absent	Typical estrogenic
Gross variegation	Yes	No	No
Pseudolobulation	Yes	Rare	Rare
Prominent ectatic vessels	Yes	Rare	Rare
Two cell types	Yes	No	Only in luteinized form
Hyaline plaques	No	Common	Common
Behavior	Benign	Always benign	

tumor because of the occasional presence of cells resembling signet cells. Ramzy[19] and Suarez, et al,[20] have described these tumors as signet - ring stromal tumors of the ovary. However, the cytoplasm of SST cells does not contain PAS or mucin positive material and the nuclei appear benign.[1,2,14-16]

In conclusion, SST is an uncommon benign stromal ovarian tumor occurring predominantly in women below 30 years of age. The characteristic histologic features are pseudolobulation of cellular zones separated by broad swathes of acellular collagenous or edematous connective tissue with a hemangiopericytomatous pattern of thin walled - dilated blood vessels in the cellular area.

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