Sclerosing stromal tumor of the ovary: A case report

E. Kuscu[1], *M.D.*; **M. Oktem**[1], *M.D.*; **H. Karahan**[1], *M.D.*; **B. Bilezikci**[2], *M.D.*; **B. Demirhan**[2], *M.D.*

[1]*Department of Obstetrics and Gynecology*
[2]*Department of Pathology, Baskent University Faculty of Medicine, Ankara (Turkey)*

Summary

Sclerozing stromal tumor of the ovary is an extremely rare neoplasm occurring predominantly in the second and third decades of life. Most patients have menstrual irregularities and pelvic pain. Infertility and endometrial pathology have also been described. A 34-year-old woman presented with hirsutism and oligomenorrhea of three months duration. Ultrasound examination showed a heterogeneous right ovarian tumor consisting of predominantly solid tissue with several loculated cysts. On T_2-weighted pelvic MR images, signal intensities of the cystic components were high and those of the solid components were heterogeneous, ranging from intermediate-high to high. Dynamic MRI marked early enhancement of solid components in the right ovary. The specimen obtained from endometrial curettage showed proliferative endometrium. Preoperative serum levels of tumor markers were in normal range: preoperative serum levels of testosterone (T) (2.42 ng/ml; normal for adult females 0.1-0.8 ng/ml) and dehydroepiandrosterone-sulphate (DHEA-S) (232.4 μg/dL; normal for adult female, 35-430 μg/dL) were measured and the T value was found increased. At laparotomy, a left ovarian mass was found attached to the right infundibulopelvic ligament and a left oophorectomy was performed. The mass was described as benign by frozen analysis. Definitive histopathological diagnosis was sclerozing stromal tumor of the ovary (SST). The histologic features included a pseudolobular pattern with focal areas of sclerosis and a two-cell population of spindled and polygonal cells. Immunohistochemical studies showed positive smooth muscle actin and negative cytokeratin, keratin, S100 and desmin. The T value decreased postoperatively (0.57 ng/ml).

Key words: Sclerosing stromal tumor; Ovary.

Introduction

Sclerosing stromal tumor of the ovary (SST) is an uncommon neoplasm that is histologically and clinically distinct from fibroma and thecoma. Chalvardjian and Scully first reported SST as a distinct entity in 1973 [1]. The distinguishing histological features of this tumor are increased vascularity, prominent focal sclerosis, and cellular patterns of pseudolobulation separated by edematous connective tissue. Clinically, this neoplasm is typically found in younger women, with 80% of cases diagnosed before age 30. This feature is unique to this particular ovarian stromal tumor. All the SSTs reported to date have been clinically benign, and all but one has been unilateral. The most common signs and symptoms are menstrual irregularity and pelvic pain. The majority of diagnosed SSTs have been non-functional, but some have produced both estrogenic and androgenic hormones. Currently, there is no known specific marker for this type of tumor [2].

We describe a case of a woman with SST who presented with hirsutism, oligomenorrhea, and a palpable right adnexal mass.

Case Report

A 34-year-old, gravida 2, para 2, woman presented with moderate hirsutism and oligomenorrhea of three months' duration. Her medical history included nine months of treatment with oral contraceptives and cyproterone acetate. The hirsutism progressed after these drugs were discontinued. Physical examination revealed hair growth on the patient's chin and neck, and a male suprapubic hair pattern. A solid, mobile, mass was palpated in the area of the right ovary. Transvaginal ultrasonography showed what appeared to be a 97 x 72 x 91 mm complex right ovarian tumor consisting of predominantly solid tissue and several loculated cysts. T2-weighted pelvic magnetic resonance imaging (MRI), revealed high signal intensity in the cystic components and heterogeneous signal intensity (intermediate-high to high) in the solid components. Dynamic MRI showed marked early enhancement of the solid components of the right ovary. An endometrial specimen obtained by curettage showed proliferative endometrium. The patient's preoperative serum levels of tumor marker cancer antigen (CA) 125 (32.4 U/ml), CA 19-9 (2.5 U/ml), alpha-fetoprotein (4.2 ng/ml), and ß-human chorionic gonadotropin (0 mIU/ml), and were all in the normal range. Measurement of serum dehydroepiandrosterone-sulfate (DHEA-S) and testosterone (T) levels preoperatively showed that T was elevated (Table 1).

At laparotomy, we found a yellow, smooth-surfaced, lobulated left ovarian mass attached to the right infundibulopelvic ligament. The patient's uterus, right ovary, and other pelvic organs were all normal. Peritoneal cytologic sampling showed no atypical cells. Left oophorectomy was performed and frozen-section analysis identified the mass as benign, so the operation was completed. The postoperative recovery was uneventful, and the definitive histopathological diagnosis was SST. Follow-up testing showed that the patient's T value normalized after surgery.

Pathological findings: Gross examination revealed a well-encapsulated 12.5 x 10 x 7.5 cm mass with a smooth bosselated surface. Sectioning showed that the tumor was mainly solid with some cystic areas. These solid and cystic portions were white- to orange-colored and exhibited hemorrhagic foci. Microscopic examination revealed tumor cells arranged in a

Revised manuscript accepted for publication February 20, 2003

Table 1. — *Serum levels of testosterone (T) and dehydroepiandrosterone-sulfate (DHEA-S) pre- and postoperatively.*

	Preoperative	Postoperative
T (ng/ml)*	2.42	0.57
DHEA-S (μg/dL)**	232.4	229

*Normal level for adult females, 0.1-0.8 ng/ml; **Normal level for adult females, 35-430 μg/dL.

pattern of pseudolobules separated by stromal edema and hyalinized tissue. The mass showed prominent vascularity and contained two main types of tumor cells, namely, spindle cells and round-polygonal cells with clear vacuolated cytoplasm. The pseudolobulated tumor cell pattern appeared similar to that seen in hemangiopericytomas. Each cell was surrounded by silver-stained reticulin fibers. Immunohistochemical analysis showed that the tumor cells were negative for cytokeratin, keratin, S100 and desmin, but positive for smooth muscle actin (Figures 1-3).

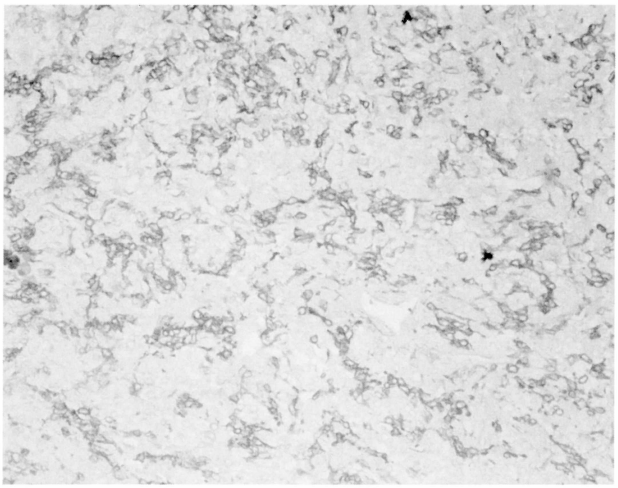

Figure 1. — Smooth muscle actin positivity (H&E, x 20).

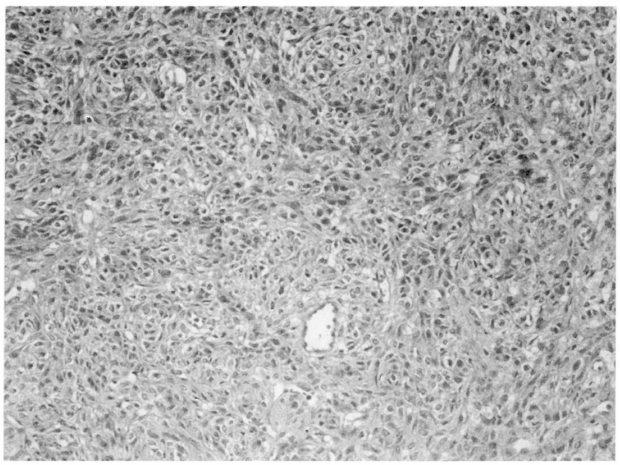

Figure 2. — Cystic and cellular areas of the tumor containing spindle cells and round-polygonal cells with clear vacuolated cytoplasm (H&E, x 20).

Discussion

Sclerosing stromal tumor of the ovary is a rare neoplasm that is most often diagnosed in the second and third decades of life. The average age of women with these tumors is 21 years. Most SSTs develop in the right ovary, but one case report describes an SST in an accessory ovary [3, 4]. The main histological features of these masses include a pseudolobular cell pattern with focal areas of sclerosis, prominent vascularity, and a two-cell population of spindle cells and polygonal cells. Most SSTs do not recur after oophorectomy or conservative resection. Our patient was 34 years old. At surgery, we found a left ovarian mass attached to her right infundibulopelvic ligament. The treatment was oophorectomy. The histological features of the specimen were similar to those detailed above.

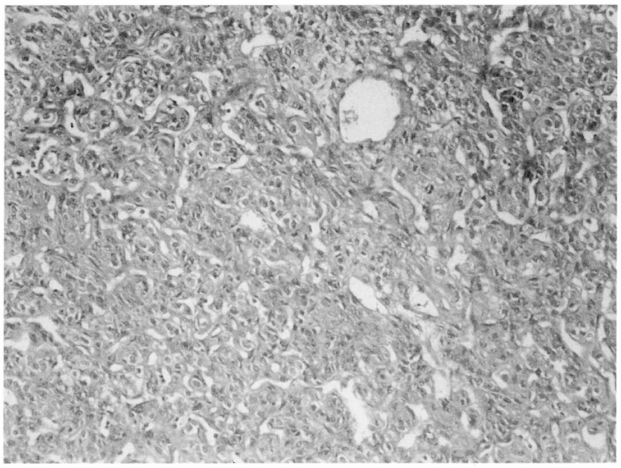

Figure 3. — The hemangiopericytoma-like pattern of tumor cells that is characteristic of sclerosing stromal tumor of the ovary (H&E, x 20).

The most common symptoms in patients with SSTs are pelvic pain and irregular menses, but many cases are asymptomatic. In general, most investigators consider these tumors to be endocrinologically inactive. However, steroidogenic activity has been clinically demonstrated in a number of instances; postmenopausal bleeding, endometrial hyperplasia and adenocarcinoma may be associated with SST [5-7]. These neoplasms can also cause marked maternal virilization during pregnancy [8-10]. Recent investigation has shown that some SSTs test positive for vascular endothelial growth factor [11, 12]. Our patient presented with hirsutism and prolonged menstruation of three months' duration. Endometrial sampling showed proliferative endometrium. Preoperative analysis of T level revealed a high value. After surgery, the T level dropped markedly. All these findings suggest that our patient's tumor was hormonally active. As mentioned above, there is currently no specific marker for SST. In our case, testing showed low levels of all tumor markers in the panel we assessed.

In the vast majority of cases, it is not possible to diagnose this uncommon ovarian tumor on the basis of preoperative clinical and sonographic findings. The physician might suspect a SST if a young woman (usually younger than 30 years) presents with a unilateral adnexal mass (5 cm or larger) that is predominantly solid on ultra-

sonography. However, evaluating serum hormone levels and all tumor markers in an attempt to reveal the hormonal activity of the lesion and correlate this with clinical findings is neither justified nor cost-effective.

Conclusion

Sclerosing stromal tumors of the ovary are rare, benign, unilateral neoplasms that are typically diagnosed in women younger than 30 years of age. More cases must be analyzed in order to comment with any certainty on the steroidogenic activity of these masses.

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Address reprint requests to:
M. OKTEM, M.D.
Onur Sokak, 38/9
06570 Maltepe Ankara (Turkey)