

CLINICAL CASE

Sclerosing stromal tumor of the ovary: report of three cases

E.U. Akyildiz¹, S. Ilvan², Z. Calay², H. Oktar³

¹State Institute of Forensic Medicine, Istanbul; ²Departments of Pathology and ³Histology, Istanbul University, Cerrahpasa School of Medicine, Istanbul, Turkey

Summary

We present 3 cases displaying sclerosing stromal tumor (SST) of the ovary and describe their immunohistochemical and electron microscopic findings. The patients were 23, 24 and 28-year-old and had an ovarian mass. Histolog-

ically, the tumors had the typical appearance of SST of the ovary. A pseudolobular pattern which was composed of cellular areas and less cellular areas was seen on low-power microscopy.

Key words: diagnosis, ovary, sclerosing stromal tumor

Introduction

Ovarian sclerosing stromal tumor is a distinctive type of benign ovarian tumor. It is a rare, usually unilateral tumor, which was initially described by Chalvardjian and Scully in 1973 [1-5]. SST predominantly affects young women with a mean age of 27 years [6]. The most common clinical symptom related to SST is menstrual irregularity. Some of these tumors have been found to show increased estrogen and/or androgen secretion. In differential diagnosis, thecoma, fibroma and hemangiopericytoma must be considered [2].

We report on 3 cases with SST of the ovary and describe their immunohistochemical and electron microscopic findings.

Case presentations

Case 1

This patient was a 28-year-old woman describing pain in the pelvis, with a left ovarian mass. The tumor was resected and macroscopically the left ovary was almost completely replaced by a mass measuring 5×4×2 cm. The tumor had a smooth, yellow-dirty white cut surfaces; there were also small cysts up to 0.5 cm in diameter.

Case 2

A 24-year-old woman was admitted in the hospital with prolonged menstrual bleeding. Preoperative diagnostic workup revealed a left ovarian mass. After tumor resection, the macroscopic appearance of the surgical specimen showed that the left ovary was replaced by a mass measuring 9×5.5×5 cm. The tumor was dirty white-purple in colour and on the cut surface there were multiple cysts filled with serous fluid.

Case 3

This patient was a 23-year-old woman complaining of pain in the pelvis. Physical examination and abdominal ultrasonography revealed a left ovarian

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Author and address for correspondence:

Dr. Elif Ulker Akyildiz
Adli Tip Kurumu
Esekapi
Istanbul
Turkey
Tel: +90 212 5850660-148
Fax: +90 212 5307451
E-mail: ulker33@yahoo.com

mass. The macroscopic appearance of the removed tumor showed a left ovarian mass with smooth surface, measuring 10×10×7 cm. Section surfaces revealed a central cystic space 5 cm in diameter, filled with blood and mucinous material (Figure 1).

Histology

Histologically the tumors had the typical appearance of SST of the ovary. On low-power microscopy a pseudolobular pattern was seen, which was composed of cellular areas and less cellular areas. Ectatic blood vessels were seen in cellular areas (Figure 2). On high-power microscopy 2 cell types were intermingled within the cellular areas (Figure 3). One type was with round to polygonal cells with clear cytoplasm, and the other type was with spindle-shaped cells. Some of the polygonal cells assumed a signet

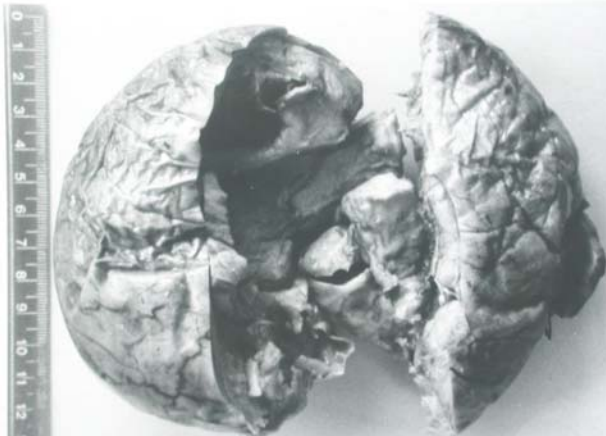


Figure 1. Case 3. Cut surface of the ovarian mass with a central cystic space 5 cm in diameter.

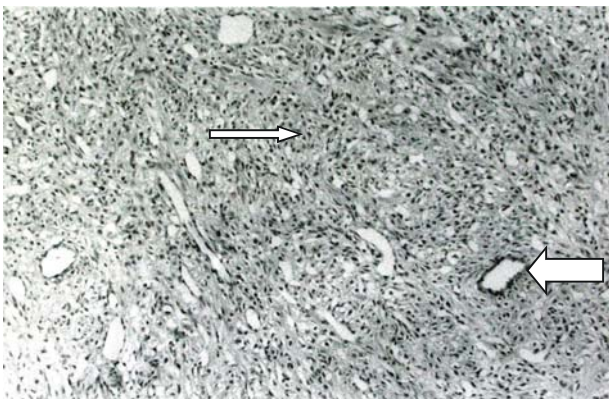


Figure 2. Two cell types intermingled within the cellular areas (thin arrow) and ectatic blood vessels (thick arrow) (H&E ×100).

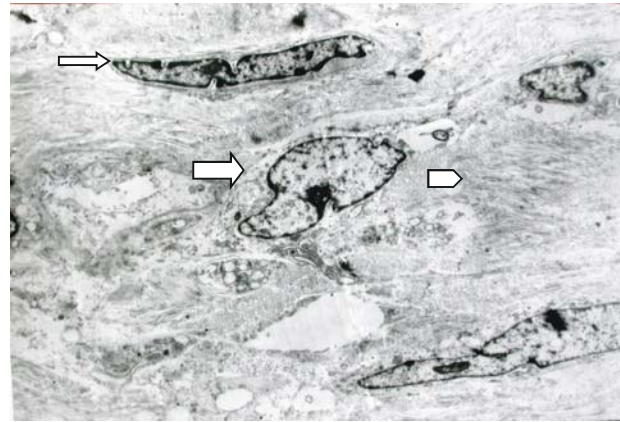


Figure 3. Electron micrograph from a representative area of the tumor. There are 2 cell types: the fibroblast-like cell with elongated nucleus (thin arrow) and the cell with round-oval nucleus (thick arrow). Collagen fibers lie around the cells (arrowhead) (lead citrate and uranyl acetate stain ×2000).

ring configuration. Poorly cellular areas were collagenous and edematous. Mitotic figures were absent. Residual ovarian tissue was not apparent on microscopic examination.

Immunohistochemically the tumor cells were positive for vimentin and desmin. Some tumor cells showed immunohistochemical staining for smooth muscle actin (SMA).

Ultrastructurally, the tumor was composed of two cell types. One type resembled a fibroblast with elongated nucleus and spindle-shaped cytoplasm. The other cell type had a round or ovoid nucleus and abundant clear cytoplasm. Collagen was present around the tumor cells.

Discussion

SST is a rare, benign, usually unilateral ovarian tumor with a prevalence of 1.5 to 6% among ovarian stromal tumors [7,8]. In 1973, Chalvardjian and Scully [1] reported 10 cases of a benign ovarian tumor and they chose the term “sclerosing stromal tumor” for these neoplasms. Unlike the other stromal tumors (thecomas and fibromas) which tend to occur in the 5th and 6th decade of life, more than 80% of SST have been encountered during the 2nd and 3rd decade of life with an average age at diagnosis of 27 years [6,9]. Fefferman et al. [6] reported a case of SST of the ovary in a premenarchal female. Our patients were 23, 24 and 28 year-old.

SST is usually an unilateral tumor. Korobowicz et al. [7] reported 3 cases, including one bilateral case. The tumors in our cases were unilateral.

Section surfaces of SST are solid and white, but they often show areas of edema and cyst formation. SST can rarely be seen as unilocular cyst [9]. In our cases there were cystic spaces on tumor cut surfaces.

Characteristic for SST is a pseudolobular pattern, with cellular nodules separated by less cellular areas of densely collagenous or edematous connective tissue, sclerosis within the nodules, prominent thin-walled vessels in some of the nodules and a disorganized admixture of fibroblasts and rounded vacuolated cells within the nodules [9].

Many authors emphasize that SST differ from fibroma, thecoma and lipoid cell tumor, both clinically and pathologically. Saitoh et al. [4] performed an immunohistochemical analysis in 2 SST cases and found that epithelial markers such as keratin and epithelial membrane antigen (EMA) were negative, but vimentin and desmin were positive. Additionally they showed that SMA was focally positive. In contrast, fibrothecomas were negative for desmin and SMA. Stylianidou et al. [10] presented a young woman with infertility and irregular menses, and demonstrated predominant positivity for SMA and consistent positivity for laminin and vimentin. In our cases the tumor cells were positive for desmin, and cells, especially those located in perivascular areas, were positive for vimentin, while some tumor cells showed immunohistochemical staining for SMA.

SST must be differentiated also from massive edema of the ovary which occurs predominantly in the younger age group. Additionally, ovarian structures such as follicles and corpora lutea or albicantia can be identified on microscopic examination of massive edema [1].

Krukenberg tumor must be considered in differential diagnosis, due to the occasional presence of cells resembling signet ring cells in SST. However, SST cells do not contain PAS-positive material [1].

Saitoh et al. [4] investigated 2 cases of SST of the ovary with electron microscopy and demonstrated the existence of smooth muscle-like cells. They emphasized that SST is derived from pluripotential cortical stromal cells differentiating toward a variety of cells such as theca interna and externa, endothelial cells, smooth muscle cells, fibroblastic cells and mature stro-

mal cells. In our cases we observed two cell types: fibroblast-like cells with elongated nucleus and cells with round-oval nucleus, which originated from ovarian stroma.

In conclusion, the morphological, immunohistochemical and electron microscopic findings of our 3 cases were consistent with SST cases reported in the relevant literature.

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