Eastern Journal of Medicine 19 (2014) 51-53

Case Report

Sclerosing stromal tumor of the ovary with torsion in a postmenopausal woman with elevated serum CA-125 levels

Gülşah Balık[a], Şenol Şentürk[a,*], Işık Üstüner[a], Mehmet Kağıtcı[a], Cüneyt Yurdakul[b], Emine Seda Güvendağ Güven[a]

[a]*Department of Obstetrics and Gynecology, Faculty of Medicine, Recep Tayyip Erdoğan University, Rize, Turkey*
[b]*Department of Pathology, Faculty of Medicine, Recep Tayyip Erdoğan University, Rize, Turkey*

Abstract. Sclerosing stromal tumor of the ovary is a subtype of sex cord stromal tumors which is hormonally inactive, benign and extremely rare. Most of the patients are below 30 years of age. These tumors do not cause an increase in tumor markers levels such as CA-125. In here, we present a 66-year-old postmenopausal woman with acute abdomen, elevated CA-125 level and torsion of sclerosing stromal tumor of the ovary.

Key words: Sclerosing stromal tumor, ovary, CA-125

1. Introduction

Sclerosing stromal tumor (SST) of the ovary is a rare subtype of sex cord stromal tumors of the ovary. These tumors are benign, usually seen unilaterally in ovary of young patients, but postmenopausal presentation is rare (1). SST infrequently leads clinical symptoms. The most common clinic symptom is abdominal pain which occurs when the tumor fills the pelvic cavity (2). A few hormonally active cases have been reported in literature (3,4), but these tumors are typically hormonally inactive (2). Thecoma, fibroma, other sex cord stromal tumors, epithelial ovarian tumors and Krukenberg tumors should be considered in the differential diagnosis (1,2).

2. Case report

A 66-year-old postmenopausal woman was admitted to our outpatient clinic with a history of abdominal pain and dyspeptic complaints for one month. On examination, there was a mass which filled up the entire pelvic cavity. Abdominal ultrasonography showed a 16.5 cm × 13.6 cm × 10.8 cm solid mass in the right adnexal region with focal cysts inside the tumor. A large volume of ascitic fluid was also visible. Alpha-fetoprotein, human chorionic gonadotropin (hCG), cancer antigen (CA) 15-3, CA 19-9 levels were within normal range but CA-125 level was 160 IU/mL. Contrast-enhanced computed tomography scan showed 15 cm × 14 cm × 10 cm, predominantly solid, heterogeneous, rounded pelvic mass originating from posterior side of the uterus and reaching up to the level of umbilicus. Endometrial biopsy was performed and an operation was planned. The patient was admitted to emergency service with severe abdominal pain 3 days later. Rebound tenderness and stiff abdomen were detected on physical examination. The laboratory findings were as followings: CA-125: 239 IU/mL, WBC: 29800 cells/mm^3, CRP: 32.7 mg/L, ESR: 110 mm/hr. Blood and urine cultures were taken and a broad-spectrum antibiotic therapy was initiated. There was no growth in blood culture, but urine culture showed growth of Enterococcus spp. The WBC level decreased to 19100 cells/mm^3 on the 2nd day of antibiotic therapy. A laparotomy was performed and a pelvic mass with torsion entangled for 4 times originating from the right ovary was detected. The tumor was soft and hemorrhagic. It could not be distinguished from

*Correspondence: Şenol Şentürk, MD,
Department of Obstetrics and Gynecology, Faculty of Medicine, Recep Tayyip Erdoğan University, İslampaşa Mahallesi, Şehitler Caddesi,
No.74, 53000 Rize/Turkey
Telephone: +90 464 2170370
Fax: +90 464 2170364
E-mail: dr.senturk@hotmail.com
Received: 10.05.2013
Accepted: 14.07.2013

the ovary. Right salpingo-oophorectomy was performed and a SST was detected by frozen section. Hysterectomy and left salpingo-oophorectomy were performed. The patient was discharged on the 4th postoperative day with no complication. The postoperative follow-up was uneventful.

Macroscopic examination showed a grey-creamy colored, soft tumor on the cut surface of the right ovary with hemorrhagic appearance and 14x13 cm in size. There were 5 cystic lesions 5 mm in sizes which were filled up with serous fluid on the cut surface of the left ovary. Endometrial thickness was normal. Subserous myomas 2x2 cm and 1x1 cm in size were also noted on the uterus (Figure 1).

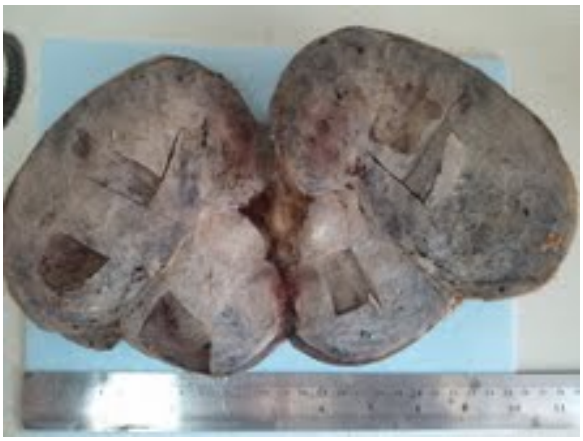

Fig. 1. Gross examination of the sclerosing stromal tumor reveals a primarily solid tumor with grayish-white and yellow nodular areas with edematous change.

Microscopic examination showed pseudolobular, fibroblastic spindle cells and there were round shaped vacuolized cells. A benign tumor constituting cellular pseudolobules separated by hypocellular/sclerotic connective tissue and a large number of thin-walled vascular structures within the pseudolobules were observed (Figure 2).

Immunohistochemical examination showed that the cells were weakly positive for estrogen receptor, weakly positive for progesterone receptor and positive for smooth muscle actin (SMA) (Figure 3). Vimentin, desmin, inhibin, calretinine, S-100 and Pan-CK were negative and Ki-67 was 1-2%. The diagnosis of SST of the ovary was made with these findings.

3. Discussion

SST of the ovary was first described as a subgroup of the sex cord stromal tumors by Chalvardjian and Scully in 1973 (1). SST is commonly seen in the second or third decades of life, but a few postmenopausal cases have been reported in literature such as this case (2).

Menstrual irregularities can occur in the reproductive ages, but SST can also be asymptomatic. Sclerosing tumors originate from ovarian stroma. Thus hormonal activity is unexpected. However, there are a few juvenile cases which have been reported with androgenic hyperactivity in the literature (3,4). Weak estrogen and progesterone receptor positivity were detected with immunohistochemical examination in our case.

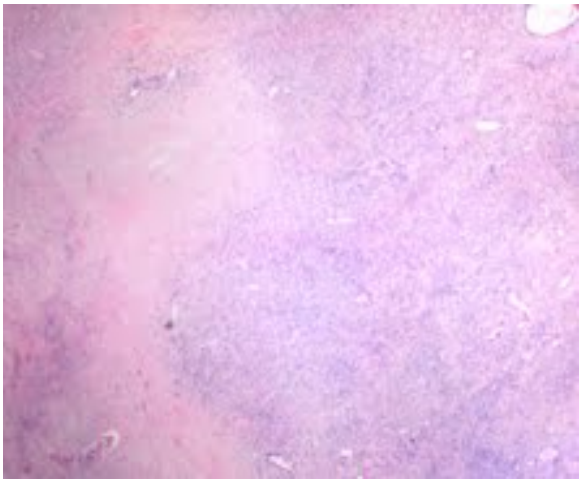

Fig. 2. Pseudolobular pattern with cellular nodules separated by less cellularly dense fibrous or edematous areas and hemangiopericytoma-like branching vessels [hematoxylin and eosin (HE) stain ×40].

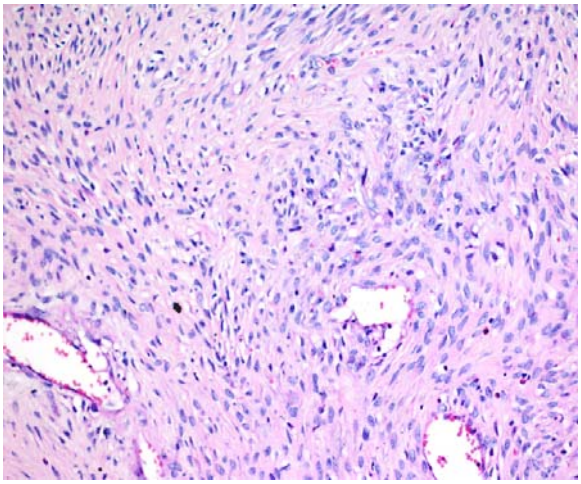

Fig. 3. Cellular nodules composed of fibroblast-like cells and rounded vacuolated cells (HE stain ×400). Tumor cells were immunopositive for α-inhibin.

SSTs have solid and cystic appearance and they look like malign tumors. The diagnosis can be made with histopathological and immunohistochemical examinations. Sclerosis, vascularization, collagenization and heterogenity (pseudolobulation, hypocellularity and hypercellularity) can be identified in histopathological examination. Existence of pseudolobulation and vascularity is important in making differential diagnosis from fibromas and thecomas. Additionally, fibromas and thecomas are usually seen in advanced ages in contrast to SST (5).

Specific immunohistochemical tests such as inhibin and calretinin are generally used in the differential diagnosis of sex cord stromal tumors (6). Granulosa cell tumors are a subtype of sex cord stromal tumors in which 100% staining for CD 99 and inhibin are expected. SMA is another antibody which is used for the diagnosis of tumors originating from theca externa. Desmin and SMA are used for differential diagnosis of fibrothecoma. Positive staining for vimentin, calretinin, CD 99 and inhibin, absence of mitotic activity and nuclear atypia, low positivity in Ki67 proliferation index and negativity of epithelial factors and S100 are expected in SST (6, 7). Low positivity in Ki67 proliferation index (1-2%) and SMA positivity were detected in our case. Krukenberg tumor should be considered in the differential diagnosis of SST (2). An immunohistochemical examination is not enough to identify SST, but also macroscopic and microscopic features can be helpful for making the diagnosis (2,8). The study performed by Zekioglu et al. (9) had showed the followings: vimentin 14/14 cases positive; smooth muscle actin, 14/14 cases positive; desmin, 14/14 cases positive; CD 99, 4/14 cases positive; inhibin-α, 14/14 cases positive; estrogen receptor, 0/14 cases positive and progesterone receptor, 2/14 cases positive. Although the present case showed an abnormal CA-125 value, immunohistochemical staining by the CA-125 antibody was negative. Increase of tumor markers are not expected in SST. CA-125 level of the case was 160 IU/mL in the initial evaluation and it increased to 269 IU/mL on the third day. Severe abdominal pain, tenderness in deep palpation and leukocytosis were detected on examination which was made 3 days after the first examination. It was considered that increase in CA-125 levels was a result of peritoneal irritation which was caused by pelvic mass. In addition abnormal elevation of CA 125 plasma levels in patients with adnexal masses may be correlated with episodes of partial ovarian torsion (10). CA-125 level decreased to the normal range after the operation.

The treatment of the SST is oophorectomy and surgical excision. SST usually occurs in the reproductive ages so intraoperative diagnosis is important to preserve fertility. These tumors should be considered in frozen section examination of the patients who desire further fertility and undergo surgery for an adnexal mass. Fertility preserving surgery must be the first line treatment option for this kind of tumors (2). There have been no recurrences reported in the literature in patients who have undergone only oophorectomy and surgical excision.

As a result, in the follow-up of the postmenopausal pelvic mass, physicians should consider the torsion of the mass when severe abdominal pain, leukocytosis, peritoneal irritation findings and sudden increase of CA-125 levels are detected. SST should be considered in the differential diagnosis of the postmenopausal ovarian tumors.

References

1. Chalvardjian A, Scully RE. Sclerosing stromal tumors of the ovary. Cancer 1973; 31: 664-70.
2. Stylianidou A, Varras M, Akrivis C, et al. Sclerosing stromal tumor of the ovary: a case report and review of the literature. Eur J Gynaecol Oncol 2001; 22: 300-304.
3. Ho Yuen B, Robertson DI, Clement PB, Mincey EK. Sclerosing stromal tumor of the ovary. Obstet Gynecol 1982; 60: 252-256.
4. Quinn MA, Oster AO, Fortune D, Hudson B. Sclerosing stromal tumour of the ovary case report with endocrine studies. Br J Obstet Gynaecol 1981; 88: 555-558.
5. Hoskins WJ, Perez CA, Young RC, et al. Principles and Practice of Gynaecologic Oncology, Lippincot Williams and Wilkins, Philadelphia, Pa, USA, 4th edition, 2005.
6. Deavers MT, Malpica A, Liu J, Broaddus R, Silva E. Ovarian sex cord-tumors: an immunohistochemical study including a comparison of calretinin and inhibin. Mod Pathol 2003; 16: 584-590.
7. Zhao C, Vinh TN, McManus K, et al. Identification of the most sensitive and robust immunohistochemical markers in different categories of ovarian sex cord-stromal tumors. Am J Surg Pathol 2009; 33: 354-366.
8. Shaw JA, Dabbs DJ, Geisinger KR. Sclerosing stromal tumor of the ovary: an ultrastructural an immunohistochemical analysis with histogenetic considerations. Ultrastruc Pathol 1992; 16: 363-377.
9. Zekioglu O, Ozdemir N, Terek C, Ozsaran A, Dikmen Y. Clinicopathological and immunohistochemical analysis of sclerosing stromal tumours of the ovary. Arch Gynecol Obstet 2010; 282: 671-676.
10. Guerriero S, Ajossa S, Caffiero A, Mais V. Relationship between abnormally high levels of plasma CA 125 and resolution of acute pelvic pain in two women with endometrioma. Gynecol Obstet Invest 1995; 40: 61-63.