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Sclerosing stromal tumor of the ovary in a perimenopausal woman: a case report

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Summary

Sclerosing stromal tumor (SST) is a rare ovarian neoplasia deriving from the sex cord stromal tumor, which occurs usually in the second and third decades of life. However, the authors report a 46-year old multiparous woman who presented with a cystic-solid left pelvic mass, a large amount of ascites, and elevated serum CA-125, all suggesting a malignant tumor. Surgery was performed and final histopathological diagnosis of the specimen was diagnosed with SST. The authors herein report an extremely rare case of SST with a cystic-solid pelvic mass and a large amount of ascites, which is useful to demonstrate the possibility of SST in multiparous woman.

Key words: Sclerosing stromal tumor; Sex cord stromal tumor; Ovarian; Ascites.

Introduction

Sclerosing stromal tumor (SST) is an extremely rare sex cord stromal tumor of ovary and was reported by Chalvadjian and Scully in 1973 for the first time [1]. SST comprises approximately 2-6% of all ovarian sex cord stromal tumors and occurs predominantly in young women with the average age of 25 years [2]. Recently, the exact etiology of SST is still unclear. Roht *et al.* suggests that SST may develop from pre-existing ovarian theca and hormonally inactive [3]. SST is also considered to be originated from the pluripotent immature stromal cells of the ovarian cortex [4]. In an overview of all cases, until now, there are only 208 cases reported in the English literature and most of these reported cases are unilateral and well circumscribed. Bilateral SST was depicted in less than 1% of all cases [5]. Herein the authors report an extremely rare case of SST with a cystic-solid pelvic mass with a large amount of ascites.

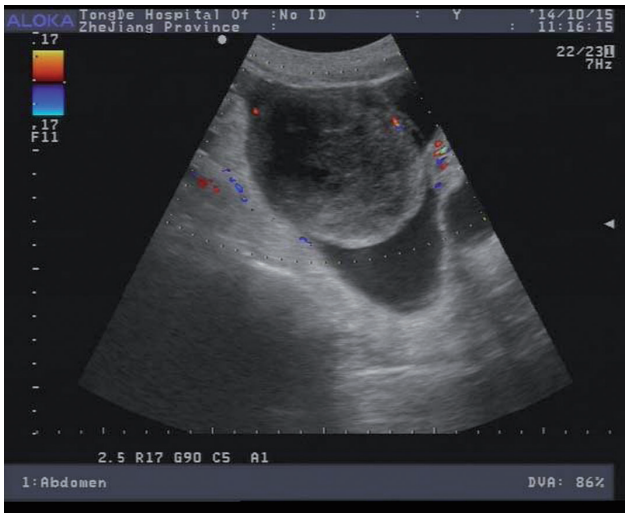

Figure 1. — Trasvaginal ultrasonography showing a 12×12 ×10 cm cystic and solid left ovarian tumor located in pelvic cavity, without exact limits, along with a large amount of ascites.

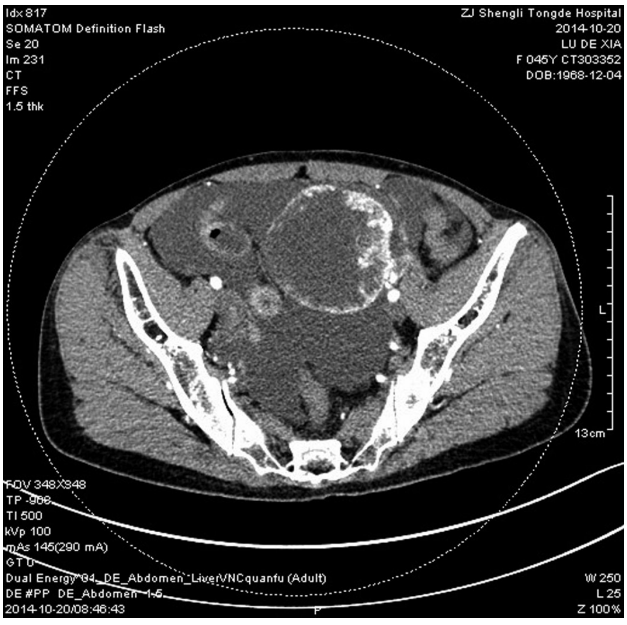

Figure 2. — Computerised tomography examination of the 12 ×12× 10.5 cm large mass in the pelvic cavity, which suggests an ovarian malignancy.

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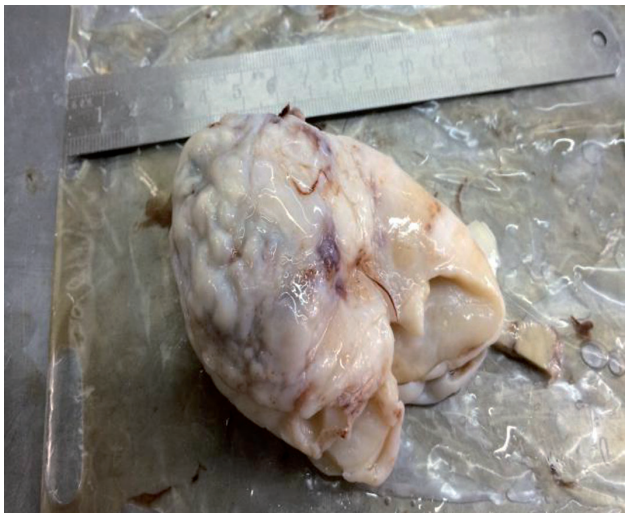

Figure 3. — The tumor in the left ovary involves its entirety, with an even surface and grayish-white color.

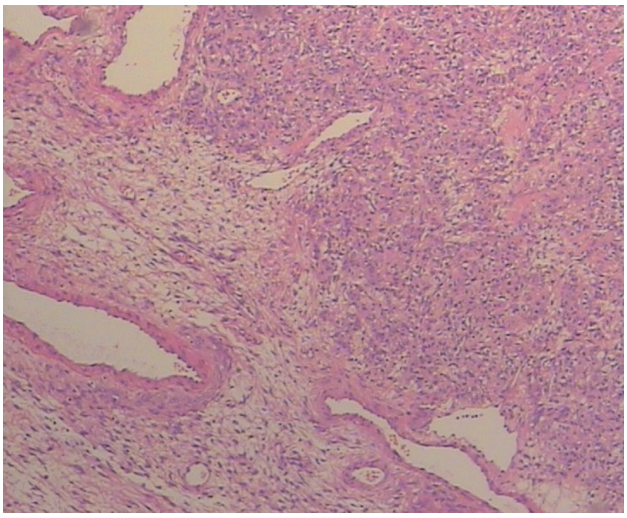

Figure 4. — Immunocytochemical staining for the pelvic mass (H&E,×40).

Case Report

In the Outpatient Department of Obstetrics and Gynecology, a 46-year-old multiparous woman complained of abdominal distension for more than a month. Physical examination showed vulva and vagina patency with no abnormality. On pelvic examination, a large pelvic mass in lower abdomen was palpable. Transvaginal ultrasonography showed a 12×12×10 cm cystic and solid left ovarian tumor located in pelvic cavity, without exact limits, along with a large amount of ascites (Figure 1). Abdominopelvic computerised tomography contrast enhanced scan showed a large inhomogeneous cystic and solid pelvic mass, which suggest a malignant ovarian tumor (Figure 2). Hence the patient was admitted to the present hospital with the diagnosis of a malignant ovarian tumor. Laboratory tests revealed an elevated serum cancer antigen-125 (CA-125) level of 119 U/ml, whereas the remaining tumor markers (including cancer antigen-199 (CA-199), carcino-embryonic antigen (CEA), alpha fetoprotein (AFP), and the routine blood tests (routine blood test, coagulation time and D-dimmer) were normal. Surgery was carried out while peritoneal cytology was also performed. On gross inspection, the uterus and right ovary were normal. Approximately 9,000 ml straw-coloured ascites and a 12×12×10 cm cystic-solid tumor on left ovary were seen in the operative. The mass was grayish-white and solid. The outer surface was uneven and well-encapsulated without no hemorrhage or necrosis (Figure 3). Frozen section was indicative of border-line sex cord stromal tumor. Therefore the authors performed a total abdominal hysterectomy with bilateral oophorectomy, peritoneal cytology, omentectomy, appendicectomy, and peritoneal biopsies. However, the final pathology disclosed an ovarian thecoma, and no malignancy cell could be found in ascites. Final immunohistochemical diagnosis showed SST (Figure 4). The patient's postoperative recovery was smooth. After surgery, the serum CA-125 declined to normal level and there was no reaccumulation of ascites. After discharged from hospital, the patient was followed up from three months until two years. Six months later, serum CA-125 level was still normal and transvaginal ultrasonography showed no ascites. The patient was found to be without recurrence.

Discussion

The common clinical manifestation of SST patients include irregular menstruations, abdominal pelvic pain or pelvic mass. In some patients, menorrhagia, infertility or masculinisation may be present. In the present case, the patient, a multiparous woman, just complained about abdominal distension, without other complaints like irregular menstruations or abdominal pain, *etc.*, which is different from the previously reported. Therefore it was difficult to diagnose SST in a 46-year-old women. Furthermore, it is almost impossible to accurately predict its character preoperatively; only clinical and ultrasonographic findings can indicate its existence [6]. On ultrasonography and computed tomography, the appearance of SST may show an increased peripheral vascularity as seen in malignant tumors. For SST, it maybe more useful in differentiating it from malignant tumors by MRI, which include a large mass with hyperintense cystic components or a heterogenous solid mass of intermediate-to-high signal intensity on T2-weighted MRI [7], unfortunately, this was not performed in the present case.

Ascites are also rare in SST [8], but in the present case, the patient had a large amount. Most SST are non-functioning tumors, which do not manipulate endocrinal function. Some cases showed elevated serum CA-125 level [9]. To the best of the present authors' knowledge, SST is considered to be a benign tumor which can be cured by oophorectomy or ovarian cystectomy, and recurrence or metastasis have not been reported. In view of the patient's age and the frozen section, the present authors performed a total abdominal hysterectomy with bilateral oophorectomy, peritoneal cytology, omentectomy, appendicectomy, and peritoneal biopsies.

Final and definitive diagnosis of SST is confirmed by histopathological and immunohistochemical assays according to the fresh specimen after surgery. SST is characterized by a dominant proportion of collagen, edematous stroma, and a large proportion of cell, which can distinguish it from fibroma, thecoma, and granular cell tumors [10]. Histologically, SST is rich in mesenchymal component and rare in epithelial component, which differs from fibroma, thecoma, and other sex cord stromal tumors [11]. Several immmunohistochemical markers of the sex cord stromal tumors had been studied in SST. After using 10% formalin fixation and paraffin embedding, immunohistochemical analysis for inhibin-α, smooth muscle actin (SMA), vimentin, calretitin, CA-125, CD99, CD177, p53, WT-1, show predominant positivity for a SMA, consistent positivity for inhibin-α, vimentin, p53, and negative for CA-125 protein, which suggest a stromal origin of the SST [12]. The characteristic vascular pattern and mitotic activity are used in the differential diagnose between SST and juvenile granulosa cell tumor [13]. CA-125, CEA, and p53 can distinguish STT and ovarian malignancy or Krukenberg tumors. PLAP and WT-1 are used to discern dysgerminoma of ovary. In the literature, inhibin-α, vimentin, calretinin, CD99, SMA, desmin, WT-1, and CD177 are reported to be useful to differentiate SST from thecoma, fibroma, and other sex-cord stromal tumors.

Because of its rarity, SST may create a diagnostic challenge. This case highlights the difficulty in discerning the diagnosis of SST from other malignancy and other sex-stromal tumor of ovary. It should be considered in the differential diagnosis in perimenopausal patients with pelvic mass, ascites, and elevated serum CA-125. The present authors believe that this case report is a useful addition to the literature to improve the accuracy of early diagnosis. This report cautions not to merely rely on clinic symptoms and neoplasms markers to differentiate benign from malignant masses.

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