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Sclerosing Stromal Tumor (SST) with Massive Blood Loss at Operation: A Case Report

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Abstract

Introduction: One of the rare ovarian neoplasms is sclerosing stromal tumor (SST). The most common age at presentation of sclerosing stromal tumor is the second and third decades of life. Usually this tumor presents with menstrual irregularity and pelvic pain as reported previously. Surgery is mandatory for diagnosis because there is not any distinctive feature to diagnose by imaging techniques.

Case Presentation: Our case in this report is a 26-year-old woman presented with pelvic pain. We conducted routine laboratory tests and checked ovarian mass tumor markers preoperatively. Due to her normal hormonal status in physical examination, we did not request more hormonal laboratory tests. However on imaging, we did not suspect benign tumor. Doppler sonography showed low resistance flow in peripheral and center of the mass. Right ovarian mass was resected and diagnosed as ovarian stromal tumor compatible with sclerosing stromal tumor. Unexpectedly at operation, we encountered severe hemorrhage from peritoneal surface so that conservative management such as packing and suturing or cauterization was not helpful. Finally, argon coagulation stopped bleeding. All coagulation laboratory tests requested by a hematologist were normal.

Conclusions: In conclusion, we believe that vascular endothelial growth factor (VEGF) production of tumor is responsible for massive bleeding.

Keywords: Sclerosing, Stromal, Ovarian Mass

1. Introduction

Sclerosing stromal tumor (SST) is a rare ovarian neoplasm first described by Chavardjian and Scully in 1973. This tumor occurs predominantly in young women (1). Germ cell tumors occur at childhood and adolescence, and other ovarian stromal tumors and epithelial neoplasm normally happen in the 5th - 6th decade of life but sclerosing stromal tumor (SST) usually presents in the 2nd - 3rd decade of life (in younger women) (2). While in some cases palpable tumor or virilization is detected, most of the patients refer with menstrual irregularity (3). There is no particular differentiation between SST and other malignant ovarian neoplasms on ultrasonography. The most common presentation is mixed pattern including cystic and solid components. However, due to predominant vascular parts of SST, color Doppler ultrasonography of SST reveals prominent vascularity in the peripheral portion and central intercystic spaces (4). It is difficult to definitely diagnose SST, preoperatively because it often mimics a malignant tumor. Oophorectomy is usually performed in many cases of young women, and diagnosis of SST is made based on post-operative pathological examination (5). A previous study reported that vascular endothelial growth factors (VEGFs) are responsible for the vascular and edematous changes of these tumors (3). In another study, twelve cases of SST were analyzed immunohistochemically and demonstrated the expression of vascular permeability factor/ VEGF(VPF/VEGF) in the luteinized theca-like cells and the receptor in capillaries and even in medium sized blood vessels. Accordingly, the characteristic vasculature and edema of SSTs were considered to be associated with the expression of VPF/VEGF. In addition, three copies of chromosome 12 in 13% - 21% of all examined SST tumor cells were reported using fluorescence in situ hybridization (FISH) analysis (6).

2. Case Presentation

A 26-year-old woman was admitted to the oncology gynecology ward of Imam Khomeini hospital affiliated to Tehran University of Medical Sciences, and presented with

pelvic pain and menstrual irregularity for 4 months. On recto abdominal digital examination, a large pelvic mass was palpable. Vaginal examination was not carried out because she was virgin. Her hormonal status was normal. Ultrasonography showed a large solid cystic mass measured approximately 15 × 20 cm. Doppler sonography showed low resistance in peripheral and central portion of the mass. MRI showed multiseptated and thick wall cystic mass in the central location of pelvic cavity. Tumor markers including Alpha feto protein, lactate de hydrogenise, carsinoemberyonic antigen, beta human chorionic gonadotropin, and cancer antigen 125 levels were normal and due to normal hormonal status we did not request more serological tests. Based on radiologic opinion, despite normal tumor markers, we strongly suspected malignancy. First, the patient was admitted with the diagnosis of a malignant ovarian neoplasm. There was no significant history in her family. Routine coagulation tests were normal. There was no free fluid in abdominopelvic cavity and there was a large and float mass at right ovary. All parts of the tissue seemed to be tumoral. Thus, right ovarian mass was resected. The frozen section examination showed sex cord-stromal tumor of ovary. Unexpectedly, we noticed massive hemorrhage from pelvic peritoneal surface which could not be stopped with routine methods such as packing and electrosurgery. 4 units packed cells and 4 units fresh frozen plasma (FFP) were transfused intraoperatively. Finally, we stopped it by argon coagulator. She discharged after three days without more complications. The gross examination showed that the resected specimen consisted of creamy grayish sheet-like tissue measuring 30×13 cm with hemorrhage in some areas. The outer surface was smooth and intact. Microscopic examination showed ovarian tissue, lobular growth pattern of cellular and hypo cellular areas composed of dual cell population. Collagen producing spindle cells and lipid-containing round to oval cells were also seen. Interlobular fibrosis and marked vascularity were identified. Finally, based on the pathologic permanent report, sclerosing stromal tumor of ovary was diagnosed. After that, follow-up with physical examination and sonography every three months was recommended to her. Now, at the 9th month of follow-up, she is asymptomatic.

3. Discussion

Sclerosing stromal tumor is a rare ovarian neoplasm first described by Chalvardjian et al. in 1973 (1) Ovarian stromal tumors present in the 5th-6th decade of life but the sclerosing type usually presents in the 2nd-3rd decade of life; so, compared to other histological types, SST presents at younger ages even in pregnancy (2). While in some cases palpable tumor or virilization is detected, most patients refer with menstrual irregularity (3). Color Doppler ultrasonography of SST reveals prominent vascularity in the peripheral portion and central intercystic spaces. Angiogenesis is a particularity of this type of neoplasm (4). Ultrasound finding indicates that they are of ovarian stromal origin (7). Preoperative diagnosis is not helpful and it is difficult to definitely diagnose because it often mimics a malignant tumor. Oophorectomy is usually performed in many cases of young women, and diagnosis of SST is made based on post-operative pathological examination (5). As mentioned before, growth factors (VPF/VEGF) can be responsible for the vascular and edematous changes of these tumors (3). In a study, twelve cases of SST were analyzed immunohistochemically and demonstrated the expression of VPF/VEGF (6). SST is an uncommon type of benign ovarian sex cord-stromal tumor. As mentioned above, according to younger age of women at presentation, clinical symptoms of SST include premature menarche, menstrual irregularities, abdominal discomfort and rarely, ascites (8). The significance of these tumors is that it is necessary to distinguish the histopathology in frozen sections in order to protect the other adnexa, because of the characteristics which are observed at early ages. Our findings support that SSTs are benign tumors which stem from stroma and they are hormonally active tumors based on per clinical and immunohistochemistry (IHC) results, although there are not any specific hormonal test that can support laboratory tests such as testosterone, estradiol, inhibin, etc. (9). The characteristic histological finding of ovarian SST is the pseudo lobular pattern that is formed by the cellular nodules which are separated from each other by hypo cellular edematous and collagenous stroma. The hemangiopericytomatous pattern-like, dilated vascular structures are the characteristics of cellular areas, and the luteinized theca-like cells with vacuolated cytoplasm and fusiform fibroblast-like cells are the characteristics of hyper cellular areas. The characteristic pathological findings of the SST of ovary were observed both macroscopically and microscopically in all reported cases (10). For exact diagnosis after pre operation preparation, ultrasound imaging, and routine serologic tests, we performed laparotomy. Macroscopic evidence did not show a benign mass strongly and the ovary was tumoral universally; so oophorectomy was performed. Frozen examination reported sex cord-stromal tumor, but unexpectedly at operation, we encountered severe hemorrhage from peritoneal surface that conservative management such as packing and suturing or cauterization was not helpful. Finally, argon coagulation stopped bleeding. All coagulation laboratory tests requested by a hematologist were normal postoperatively. In conclusion, we believe that VEGF production of tumor was responsible for massive bleeding. As mentioned above, the expression of

VPF/VEGF was detected in SST by IHC examination; therefore, our concept can support that the expression of VEGF on the peritoneum surface is responsible for ongoing hemorrhage. We consulted a hematologist and requested complete coagulation profile but our findings did not show any abnormality. Now after 9 months, she does not present symptoms and her clinical examination has no significant findings and no lesion was observed on imaging study of ovaries and pelvic.

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