Different Presentations of Five Rare Cases of Sclerosing Stromal Tumor of the Ovary

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Abstract

A rare subtype of preceding neoplasm of ovary is sclerosing stromal cell tumor with few presented cases in the literature. In these case series, we describe five cases of ovarian sclerosing stromal cell tumor with different presentations in our department. Interestingly, one of our cases had elevated alfa-fetoprotein.

Keywords Sclerosing stromal tumor · Ovarian cancer · Ovary sex cord tumor · Ovary

Introduction

Ovarian sex cord tumors are counted as infrequent neoplasms compared to epithelial cell tumors by representing about 8% of all primary ovarian cancers [1]. A rare subtype of preceding neoplasm of ovary is sclerosing stromal cell tumor (SST) introduced first at 1973 [2] with distribution ranging from 1.5 to 6% [1, 3, 4]. SST, originating from the stroma of the ovary, is a benign most likely unilateral tumor seen remarkably at young ages, with 70% incidence between 14 and 29 years of age [1, 5, 6]. The most common symptoms of ovarian SSTs are menstrual irregularity and pelvic pain, and usually they are hormonally inactive solid tumors without endocrine androgenic function; however, there are some exceptions with androgenic symptoms, which are commonly occurred during pregnancy

[7–12]. Distinguishing of ovarian SST from malignant tumors may be impossible by imaging preoperatively, and it should be done using pathological examination during surgery or after surgery.

The rarity of the disease is responsible for minority of the case reports in the literature [6]. The main purpose of this research is to report five different cases of ovarian SST with different presentations.

Case Presentation

In this part, we would introduce clinical presentations, treatments, and follow-up outcomes of different five cases encountered by summarizing them in Table 1.

Based on the collected information, one patient had hirsutism attributing to androgenic effect of tumor, two had fullness of adnexa because of large size of tumor, and the two others had normal physical exam. All presented symptoms were expected based on clinicopathologic features of these tumors.

According to sonographic and postoperative evaluations, size of benign tumor ranging from 3 to 12 cm was seen. All ovarian SSTs were unilateral and seen in young women with an average age of 23.

Microscopic examination of all tumors was uniform and showed pseudolobular hypercellular areas composed of two types of cells: spindle cells fibroblast-like and round to oval cells with luteinized cytoplasm with signet-ring appearance seen in some of them. There was also a network of thin-walled hemangiopericytoma-like blood vessels. These cellular

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Table 1 Demographic data and characteristics of all cases

	Case 1	Case 2	Case 3	Case 4	Case 5
Age	23 y/o	30 y/o	27 y/o	18 y/o	17 y/o
Gravida	Null gravida	G2L1, pregnant	Single	Null gravida	Null gravida
Chief complaint	8 years primary infertility	Labor pain, term pregnancy	Protrusion of abdomen	Abnormal uterine Bleeding	Abdominal pain
Past medical history	8 years primary infertility	Absent left kidney, enlarged spleen	Nothing	2 years primary infertility	Nothing
Menstruation	Irregular	Regular	Regular	irregular	Irregular
Physical exam	Hirsutism	Nothing	Fullness of adnexa	Nothing	Fullness of adnexa
Tumor marker*	Normal	Normal	Increased α FP	Normal	Normal
Imaging	Sono:44*28 mm right ovarian mass	Sonography: pregnant uterus with 25 × 30 mm simple right ovarian cyst	Sonography: 110 × 46 mm complex mass with multiple cystic change from right ovary	Sono: 34*30 mm left ovarian mass with atypical flow (RI: 0.39)	Sono:85 × 55 × 87 mm Rt adnexal mass CT: 81 × 57 × 90 mm Rt adnexal mass MRI: large heterogeneous Rt adnexal solid mass 90 × 60 × 70 mm with degeneration
Kind of operation	Excision of right-side mass + cauterization	Cesarean section (full arrest) + resection of right ovarian mass	Right-side oophorectomy + left ovarian biopsy	Left-side ovarian cystectomy + mass resection (fertility-sparing technique)	Right-side salpingo-oophorectomy
Frozen section report	Sex cord-stromal tumor, no malignancy	Benign tumor	Sex cord-stromal tumor	SST	Benign tumor
Postop findings	Left ovarian PCO pattern + 40*30 cm right ovarian mass	30 mm right-side ovarian mass	Bilateral ovarian mass, right-side 120*40 mm	40*40 mm left-side mass with adhesion of left tube	Rt adnexal mass 80*90-mm-like dermoid cyst
Follow-up	34 months Pregnancy with letrozole after 3 months, at GA = 12 week miscarriage happened Spontaneous pregnancy 4 months later with consequence of a healthy neonate. No recurrence of tumor	30 months Not eventful during every 3 months follow-up	22 months Not eventful during every 3 months follow-up	48 months No pregnancy during 4 years. No sign of recurrence of tumor during every 3 month follow-up	26 months Not eventful during every 3 months follow-up

*Tumor marker includes: alpha-fetoprotein, CA125, HE4, CEA, CA19-9

areas were separated by edematous hypocellular fibrous tissue (Figs. 1 and 2).

Discussion

Ovarian SST, firstly reported by Chalvardjian and Scully in 1973 [2], is a very rare sex cord-stromal tumor happening predominantly in a younger age group than other types of stromal tumors [1, 5, 6]. All five reported cases were younger than 30 years with average of 23 years old. It means that the patients are in the childbearing age, for which conservative surgery of the ovaries is important for quality of life. Three patients (60%) were nulligravida, one case was single and one was pregnant. Diagnosis of ovarian SST is a challenging issue during pregnancy due to difficult differentiation of it from luteinized cells tumors [13]. The differential diagnoses are thecoma, fibroma, lipid cell tumor, ovarian granulosa cell tumor, and ovarian mucinous cystadenocarcinoma. Ovarian SST was seen not only in a normal pregnancy course with early onset severe preeclampsia but also in a young woman with ectopic pregnancy [14, 15].

Clinically, two patients had infertility and three patients represented with irregular menstrual bleeding as the most common symptoms reported in the literature [5, 6, 11, 12]. Recently, several reports have described the active ovarian SST with hormones production either estrogenic or androgenic by synthesizing dehydroepiandrosterone. Associated endometrial hyperplasia with ovarian SST may be the result of excessive hormone production by these tumors. Hence, these tumors can cause irregular menses, infertility, precocious puberty, and virilization [6].

Among the markers we tested, all had normal tumor marker except a single case with increased alpha-fetoprotein. Besides, there are only a case report of elevated CA125 [16] and a case

Fig. 1 Pseudolobular pattern with thin wall hemangiopericytoma-like blood vessels, (H&E $\times$ 100)

Fig. 2 Dual cell populations (spindle fibroblast-like cells and round cells) with luteinized cytoplasm, some with signet-ring appearance, (H&E $\times$ 400)

with elevated CA19-9 in the literature [17]; we had a case with elevated serum alpha-fetoprotein marker.

The gross appearances of SSTs of ovary mostly are solid with small cystic areas [1]. On histopathological study, the pseudolobular pattern, caused by cellular areas separated by edematous hypocellular areas and the hemangiopericytoma-like network are characteristic of ovarian SST [1, 6, 18]. The cellular areas contain dual cell population and included spindle cell producing collagen and round to oval cells with luteinized or clear cytoplasm containing lipid. The later cells are epithelioid and sometimes signet-ring like. Mitotic activity is low in this tumor [1, 6, 18]. Although immunohistochemical expression of vimentin, inhibin, calretinin, melan-A, smooth muscle actin (SMA), CD99, and CD34 are reported in SST of the ovary but epithelial markers and S100 are negative [1, 5, 6, 18]. Expression of vascular permeability factor/vascular endothelial growth factor detected on the lutein cells explains rich vascularity of the ovarian SST [18]. The differential diagnosis of SST includes the fibroma/thecoma, lipid cell tumor, vascular tumor, metastatic signet-ring carcinoma, and massive edema of ovary [1, 6, 18]. Pseudolobular pattern and prominent vascularity of ovarian SST are absent in the fibroma/thecoma, and they frequently are seen in the elderly women [1, 6, 18]. The inhibin immune reactivity can differentiate ovarian SST from vascular tumor [1]. Rarely, the vacuolated luteinized cells in SST can mimic the signet-ring cells of a Krukenberg tumor. The pathologic characteristic of Krukenberg tumor is the signet-ring cells contain mucin, immunoreactive for cytokeratin, and epithelial membrane antigen with nuclear atypia and mitotic activity. On the other hand, the vacuoles in ovarian SST cells contain lipid [6, 18].

Described sonographic features, while being nonspecific, include a mass with multinodular cystic components, irregularly thickened septa and tumor walls [19]. MRI features in two modalities of T2 and T1 are as follow: T1-weighted

images disclose intermediate signal intensity in the outer part and hypointense in the inner part of the mass. On T2-weighted images, signal intensities of the cystic components are high and those of the solid components were inhomogeneous, ranging from intermediate-high to high. Pseudolobulation of the lesion consists of low-intensity nodules set against high-intensity stroma might be seen in T2-weighted images. Dynamic MRI could display a pseudolobulated mass with peripheral contrast enhancement in the early phase [20, 21].

Treatment options of ovarian SST include fertility-sparing technique and excision of mass [22] or open or laparoscopic salpingo-oophorectomy or single oophorectomy [23]. In some centers like ours, the final decision is based on the frozen section pathology report and size of the lesion. Young age of the patients should be considered to perform less invasive surgeries.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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