

Sclerosing stromal tumor in a postmenopausal woman with an ovarian torsion

Overyan torsiyonlu postmenopozal kadında sklerozan stromal tümör

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Abstract

Sclerosing stromal tumor is a rare benign ovarian sex-cord stromal tumor, which has distinctive clinical and pathologic features. The tumors occur predominantly in the second and third decades and are histopathologically characterized by the pseudobubular pattern of the cellular and hypocellular areas with widespread areas of sclerosis and a two-cell population of spindled and polygonal cells, and marked vascularity. We presented a sclerosing stromal tumor of the left ovary in a 73-years-old patient who presented with an ovarian torsion. Although sclerosing stromal tumor of the ovary mostly occurs in young patients, some must remember sclerosing stromal tumor in older ones. Like any ovarian mass, sclerosing stromal tumor may cause the torsion of the ovary.

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Key words: Sclerosing stromal tumor; ovarian torsion

Özet

Sklerozan stromal tümör, belirgin klinik ve patolojik özelliklere sahip, nadir bir benign overyan seks-kord stromal tümördür. Büyük çoğunluğu ikinci ve üçüncü dekatta görülür ve histopatolojik olarak selüler ve sklerozisin geniş alanlarını içeren hiposelüler alanların psödobubüler paterni ve spindle/iğsi ve poligonol hücrelerden oluşan iki tip hücre popülasyonu ve belirgin vaskülarizasyon ile karakterizedir. Biz, overyan torsiyon ile başvuran 73 yaşındaki kadın hastada sol overin sklerozan stromal tümörünü sunduk. Overin sklerozan stromal tümörü çoğunlukla genç hastalarda görülmesine rağmen, bazı daha yaşlı hastalarda da sklerozan stromal tümör akla gelmelidir. Herhangi bir overyan kitle gibi, sklerozan stromal tümör overin torsiyonuna neden olabilir

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Anahtar sözcükler: Sklerozan stromal tümör, overyan torsiyon

Introduction

Sclerosing stromal tumor (SST) is a rare variant of ovarian sex-cord stromal tumor, which is first described by Chalvardjian and Scully [1] in 1973. It is a benign ovarian neoplasm occurring predominantly in young woman in the second and third decades [2-8]. Clinically it usually presents with menstrual irregularities and pelvic pain [3,6,8]. To our knowledge, all of the affected patients described in the literature were young and there is only one case presented with torsion [9]. We present a case of SST in a 73-year-old postmenopausal patient presented with an ovarian torsion.

Case

73-year-old woman presented with a left side pelvic mass, which was approximately 14 cm solid and cystic well-circumscribed mass on ultrasonographic examination. At laparotomy a brown, torsioned, and partially ruptured mass was found on the left ovary. Unilateral salpingo-oophorectomy was carried out and frozen section analysis of the mass revealed a sex-cord stromal tumor with large necrotic and hemorrhagic areas. Complementary total hysterectomy and unilateral salpingo-oophorectomy was done. The postoperative course was uneventful. The patient was discharged from the hospital after

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5 postoperative days. She has been followed on an outpatient basis without a specific finding since 12 months.

Macroscopically the tumor measured 14x11x5.5 cm, weighed 470 gr. External surface was brown and partially ruptured in gross appearance. The cut surface was hemorrhagic, brown in color and soft in consistency. On histological examination, the tissue had large necrotic and hemorrhagic areas (Figs. 1,2).

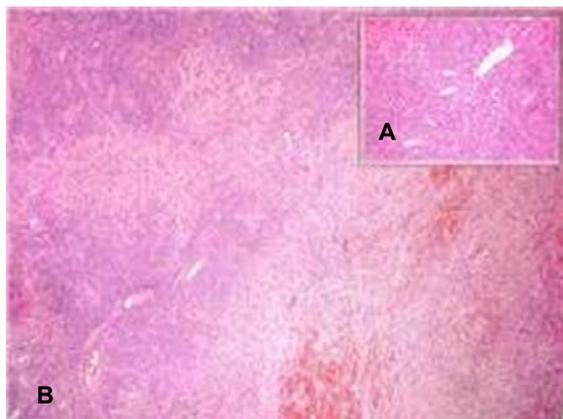


Figure 1. Pseudolobular pattern due to hypercellular and hypocellular areas with areas of hemorrhage and necrosis. The inset shows richly vascularized pseudolobule composed of spindle cells mixed with large rounded vacuolated cells (Hematoxylin and Eosin, A, x100; B, x200)

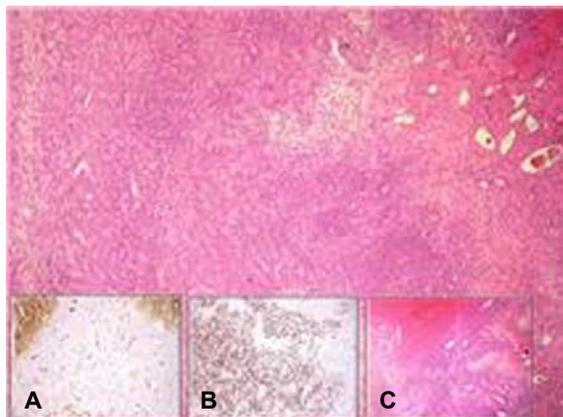


Figure 2. Immunohistochemical analysis of the tumor demonstrated positivity for estrogen and negativity for CD 34. Note the pseudolobular pattern with large areas of hemorrhage (A, CD 34, x100; B, estrogen, x100; C, Haematoxylin and Eosin, x100)

In the more preserved fields, it had a pseudo lobular appearance with cellular and hypocellular areas, and with focal hyalinizing areas (Figs. 1,2). Each tumor cell nuclei was ovoid or spindle in shape and cytoplasm was eosinophilic or clear focally. Cellular borders

were indistinguishable. Immunohistochemical analysis showed that tumor cells were positive for calretinin, inhibin, progesterone, estrogen, but negative for cytokeratin 7, CD34, vimentin, smooth muscle actin, S-100, chromogranin and sinaptophysin. The final diagnosis was sclerosing stromal tumor of the left ovary with torsion.

Discussion

Sclerosing stromal tumor is a rare benign ovarian neoplasm occurring predominantly in young woman in the second and third decades [1-8]. The oldest patient reported in the literature was 49 years-old woman [10]. 71% of tumors were found on the right side and were unilateral and benign in the literature [5,6]. Our patient was 73 years old and presented with a left side pelvic mass.

Clinically it presents with menstrual irregularities and pelvic pain [3,6,11]. USG and computed tomography findings show a complex cystic mass with marked peripheral vascularity [10,12]. Because of the cystic and solid components, they may be suspected to be malignant ovarian tumors based on US images alone, magnetic resonance imaging (MRI) and dynamic MRI may be more specific in differentiating this tumor from a malignant neoplasm and other sex-cord stromal tumors and useful for the preoperative diagnosis of SST to avoid excessive surgical intervention [11,13]. In our patient, ultrasonographic examination showed 14 cm solid and cystic well-circumscribed mass.

It is distinct from the thecoma-fibroma group and steroid cell tumors clinically, pathologically and radiologically [11]. The SST is characterized by cellular pseudolobules, prominent interlobular fibrosis, frequently marked vascularity and a dual cell population: collagen-producing spindle cells and lipid containing round or ovoid cells. The heterogeneity due to the variation in cellular size and shape are helpful features in the differential diagnosis of STT, and contrasts with the relative homogeneity of thecoma and fibromas [1-6]. On the other hand, thecomas and fibromas generally occur in the fifth or sixth decades of life [8,9].

In some cases, the differential diagnosis between SSTs and juvenile granulosa cell tumor with pronounced stromal sclerosis was difficult. However, the characteristic vascular pattern and mitotic activity were used in favor of SST, whereas tumors with follicular structures, higher mitotic activity and characteristic granulosa cell morphology were rather classified as

juvenile granulosa cell tumor [10]. Occasionally, the vacuolated cells have signet cells of a Krukenberg tumor, but the former cells contain lipid instead of mucin [8].

The etiology of SSTs is unknown. Based on the ultrastructural features, SSTs were thought to arise from pluripotent immature stromal cells of the ovarian cortex. However SSTs are proposed to be derived from a population of muscle-specific actin-positive elements from the theca externa, namely the perfollicular myoid stromal cells. On the other hand Ismail et al [14] suggested that endocrine milieu might be responsible for the morphology of SST and they may be developed from pre-existing ovarian fibromas.

In the literature inhibin, calretinin, CD34, alpha glutathione S-transferase positivity (α -GST), melan-A, müllerian-inhibiting substance, WT-1 and CD99 was reported to be useful to differentiate STT from thecoma, fibroma and other sex cord stromal tumors [14-16].

Inhibin is a specific, but less sensitive marker than calretinin in the diagnosis of ovarian sex cord-stromal tumours [17]. Also, inhibin and calretinin have been shown to be more sensitive and specific marker than CD99, A103 (melan-A), CD10 and WT-1 for ovarian sex cord stromal tumors [14, 15].

α -GST positivity within scattered cells appears to be useful in the distinction of SST from diffuse staining thecomas and no staining fibromas. CD 34 stains the endothelium of often dilated and branching vascular architecture, and clearly distinguishes SSTs from thecoma and fibromas [14].

Although many studies showed variable immunohistochemical analysis for sclerosing stromal tumors, a predominant positivity for inhibin, calretinin, smooth muscle actin and vimentin is a well-known immunohistochemical panel suggesting a stromal origin of the SST [18].

In our study, fibroblast-like cells were immunohistochemically stained for calretinin, inhibin, and negative for cytokeratin 7, CD34, S-100, chromogranin and sinaptophysin consistent with the recent literature. However, some challenging immunohistochemical findings such as positivity for progesterone, estrogen and negativity for vimentin, and smooth muscle actin showed that an immunohistochemical panel will not exactly differentiate SST from other tumours in the sex cord-stromal group. Characteristic macroscopic and histopathological features of

the tumour in addition to the patient's young age and the unilaterality of the tumour may be more important in the differential diagnosis.

Surgical removal is curative and no local or distant recurrences have been reported [3,4,11].

We presented a case of sclerosing stromal tumor in 73-year-old female and presented with left ovarian torsion. Although they mostly occur in young patients, some must remember SST in older ones. Like any ovarian mass, SST may cause the torsion of the ovary.

Foot note: This case report was presented as a poster at the 21st European Congress of Pathology in September 08-13, 2007.

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