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SCLEROSING STROMAL TUMOR OF THE OVARY

OVERİN SKLEROZAN STROMAL TÜMÖRÜ

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SUMMARY

Sclerosing stromal tumors are rare benign ovarian stromal tumors, which have distinctive clinical and pathologic features. These tumors are characterized by the pseudolobular pattern of the cellular and hypocellular areas, marked vascularity and heterogeneity of the cellular area histopathologically.

Here we described a case of sclerosing stromal tumor in a 25-years-old patient who presented with hypermenorrrhea. The histologic features included a pseudolobular pattern with widespread areas of sclerosis and a two-cell population of spindled and polygonal cells.

ÖZET

Sklerozan stromal tumor, ayrı klinik ve patolojik özelliklere sahip nadir, benign overiyal stromal tümördür. Bu tümörler histopatolojik olarak sellüler ve hiposellüler alanların psödonodüler paterni, belirgin vaskülarite ve heterojen sellüler alanlarla karakterizedir.

Bu çalışmada, hipermenore ile başvuran 25 yaşında bir hastada saptadığımız sklerozan stromal tümör olgusunu sunduk. Tanımlanan tümör dokusu yaygın sklerozis alanları, spindle ve poligonal hücreler şeklinde iki hücre populasyonu ile psödonodüler patern içeriyordu.

INTRODUCTION

Sclerosing stromal tumor (SST) is a rare, benign ovarian stromal tumor first described by Chalvardjian and Scully in 1973 (1). This tumor was classified as a distinct subtype within the ovarian sex-cord stromal tumors (2-4).

CASE REPORT

A 25-year-old woman was presented with 2-years history of hypermenorrhea. The past medical history was unremarkable.

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Physical examination and radiologic findings revealed an abdomino-pelvic mass that was suspected to be a subserosal leiomyoma. Pre-operative laboratory investigation showed that the patient's serum estradiol (E2) level was normal (118 pg/ml, ranging 40,7–242 pg/ml). She was underwent laparoscopy for definite diagnosis and optimal treatment. She was found to have a left ovarian mass measuring approximately 4 cm and removed, laparoscopically.

Grossly, a 4,5x3,5x2 cm surgical specimen presented a whitish, smooth external surface. The cut surface was white-yellow in color with orange at focal areas and was predominantly solid and glistening (Fig–1).

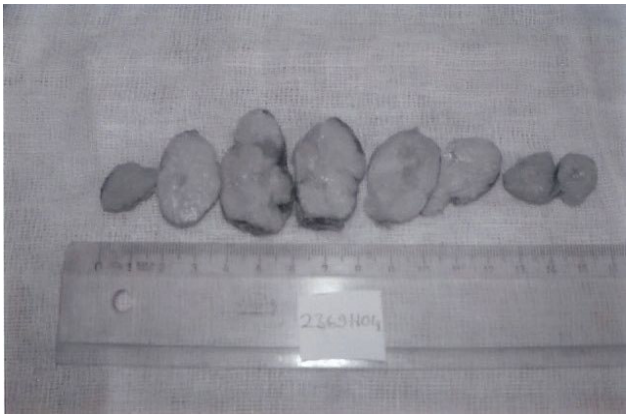

Fig-1. The cut surface of the mass.

Histologically, cellular areas separated by hypocellular areas characterized by densely collagenous, edematous, or myxoid tissue rich in vascularity creating a pseudolobular pattern were prominent (Fig-2). The cellular areas were composed of oval to spindle shaped cells, with a single oval to round nucleus and a prominent nucleolus; the cytoplasm was moderately abundant, eosinophilic, and occasioanlly vacuolated (Fig-3). Nuclear atypia and mitosis were not seen.

Fig-2. Pseudonodular pattern due to hypercellular and hypocellular areas (X40, Hematoxylen-Eosin).

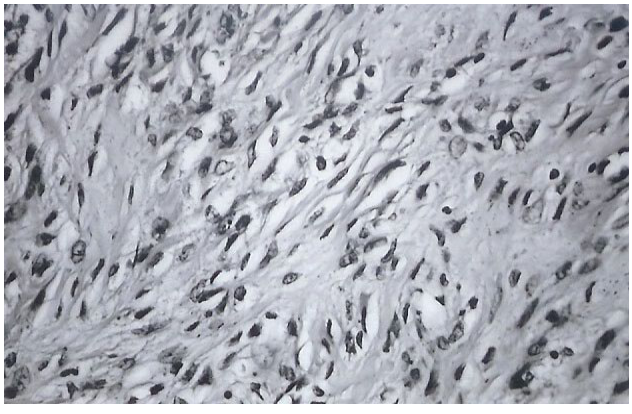

Fig-3. Oval to spindle shaped cells (X400, Hematoxylen-Eosin).

Tumor cells stained for smooth muscle actin, vimentin, progesterone, CD 34 and but not for other markers including the estrogen, pansitokeratin and desmin. In addition, CD34 staining pattern showed hemangiopericytoma-like pattern (Fig-4).

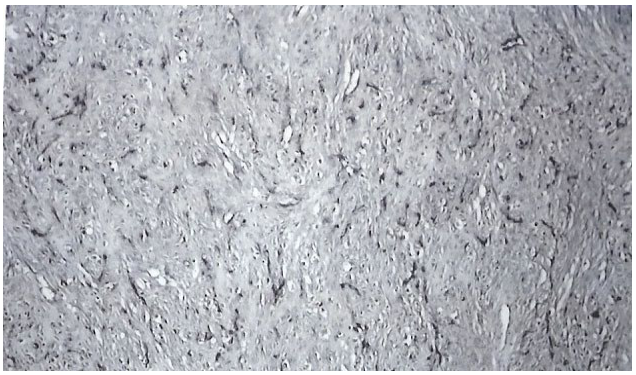

Fig-4. CD34 staining pattern showed hemangiopericytoma-like pattern (X100, CD34).

The final pathologic diagnosis was SST of the left ovary. Eight months after surgery the woman has regular menstrual cycles and no signs of recurrences.

DISCUSSION

Sclerosing stromal tumor (SST) is an extremely rare sex cord stromal tumor, occurring predominantly in the second and third decades of life (4). The most common presenting clinical symptoms in patients with SST were menstrual irregularity and pelvic pain (1,2,5). Microscopic examination reveals a heterogeneous picture that is characterized by sclerosis and a disorganized admixture of fibroblasts and vacuolated cells (6). Prominent vasculature of the nodules mimicking hemangiopricytoma is one of the features that characterize SST (5).

This tumor distincts from the fibroma and the thecoma both clinically and pathologically. More than 80 % of SSTs have been encountered during the second and third decades, whereas thecomas and fibromas are uncommon in the first three decades (2,8).

Although our tumor developed in the left ovary; 71 % of tumors were found on the right side and were unilateral and benign in the literature (5,6).

Serum levels of estradiol were normal in our patient. These findings are similar to those of other authors (6). Although 40 % of tumor cells displayed progesterone receptor positivity, estrogen receptors were not expressed in our case. Menstrual irregularity was disappeared soon after the removal of the tumor, indicating that some tumors may develop independent of stimulation by estrogen (7).

The thick peripheral rim present at US and CT histologically comprises cellular areas patterned in a nodular configuration, separated by edematous stroma with prominent vasculature (2).

Smooth muscle actin staining of the SSTs was generally marked and involved the walls of larger blood vessels and perivascular cells. The immunohistochemical identification of intracytoplasmic αGST appears to be useful in the distinction of SSTs from fibromas (9). The positive CD34 of endothelial cells highlighted the vascular structure and demonstrated clear differences between the SST and the thecomas and fibromas (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 a signet cells of a Krukenberg tumor, but the former cells contain lipid instead of mucin (8).

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

In conclusion, SST is an ovarian stromal tumor composed of heterogeneous components. The prominent vasculature, which histologically characterize SST is associated with CD34 expression.

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