Sclerosing stromal tumor of the ovary associated with Meigs' syndrome: A case report

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Summary

Sclerosing stromal tumors of the ovary are distinct, but rare benign neoplasms. These tumors appear solid and are very vascular giving the impression of malignant tumors. They occur mostly in young women. Morphologically they have distinct characteristics which differentiate them from other stromal tumors. Benign ovarian tumors associated with Meigs' syndrome are rare. In this article a case of ovarian sclerosing stromal tumor associated with Meigs' syndrome in a 17-year-old women is described and the differential diagnosis is also discussed.

Key words: Sclerosing stromal tumor; Ovary; Meigs' syndrome.

Introduction

In 1973, Chalvardjian and Scully, described sclerosing stromal tumor (SST) of the ovary as a rare benign tumor separate from other ovarian stromal tumors [1]. To our knowledge, fewer than 100 cases have been reported in the English language literature. More than 80% of SSTs occur before the age of 30 and all reported cases have been benign [2-4].

In this article a case of sclerosing stromal tumor of the ovary with ascites is described together with a review of the literature.

Case Report

A 17-year-old girl with pelvic pain and irregular menses was referred to our hospital. Her physical examination revealed a huge, non-tender pelvic mass and normal female sex characteristics. She had had irregular menstrual cycles since her menarche at age 14. Abdominal X-ray showed midline pelvic soft tissue density. Ultrasonography revealed a moderate amount of ascites and a solid mass of 20.5 x 10.9 cm in diameter in the right adnexal region. Her laboratory tests were unremarkable and tumor markers including α-fetoprotein, carcinoembryonic antigen and CA 19-9 were all within normal limits except for an elevated serum level of CA 125 (193 U/ml). A huge, greyish-white, ovoid mass with a smooth surface involving the right ovary was shown at exploratory laparatomy. Her left ovary, uterus and other pelvic and abdominal organs were grossly normal without any impression of metastasis or invasion. There was approximately 400 cc clear fluid in the pelvis. A right salpingo-oophorectomy was performed and the specimen was submitted for frozen section and evaluated as non-malignant (sclerosing stromal tumor?). The postoperative course was unremarkable.

On gross examination, the right ovary was enlarged, measuring 25 x 18 x 15 cm, weighing 970 g, and with a smooth surface. On cross section, it was almost entirely replaced by a grayish-white, firm, rubbery tumor with a focal gelatinous and cystic appearance.

Microscopically, the tumor showed a pseudolobular pattern with cellular areas separated by edematous and collagenous areas (Figure 1). The cellular areas consisted of round cells with clear cytoplasm, spindle-shaped cells with collagen fibres and well developed vascularity (Figure 2). The mitotic index was 3 mitoses per 10 high-power fields. A reticulin stain showed numerous fibers surrounding individual cells. An oil Red O stain demonstrated abundant intracellular fat. No Reinke crytals or Call-Exner bodies were identified. Normal ovarian stroma was not observed. Immunohistochemical analysis demonstrated positivity for vimentin and smooth muscle actin in the cellular areas of the tumor. The pathologic diagnosis was sclerosing stromal tumor of the ovary.

The patient has been free of disease for two years since the diagnosis.

Discussion

SSTs have distinct clinical and pathologic features that warrant their differential diagnosis from other ovarian stromal tumors. They occur in a younger age group, with an average age of 28 years at diagnosis. Whereas fibroma and thecoma are uncommon in the first three decades of life, 80% of SSTs have been diagnosed during this period. Immunohistochemistry of desmin and smooth muscle actin is useful in distinguishing SSTs from thecofibroma [3]. It has been suggested that this rare tumor is derived from a population of muscle-specific actin positive elements from the theca externa, namely the perifollicular myoid stromal cell [5].

The most common presenting symptoms are menstrual irregularities and pelvic pain as in the presented case. Most of the reported cases of SSTs are nonfunctional, however some of them may produce estrogenic or androgenic hormones that cause anovulatory irregular menstrual bleeding [6]. Surgical removal of SSTs was

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Figure 1. — Low-power microphotograph demonstrating a pseudolobular pattern of tumor (H&E x 40).

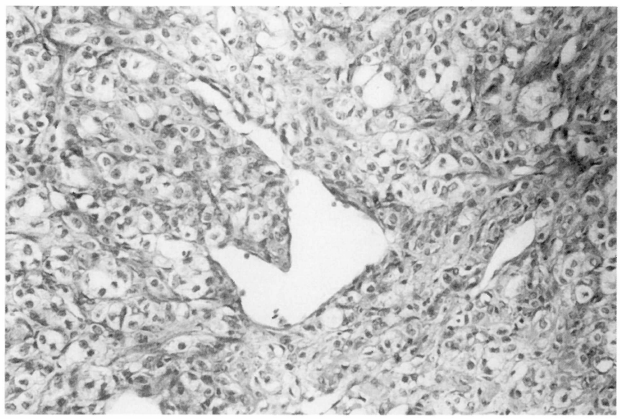

Figure 2. — A cellular area is shown around cells with clear cytoplasm, spindle-shaped cells, dense collagen formation and well developed vascularity (H&E x 200).

reported to cause ovulatory cycles to be resumed in several patients, one of whom became pregnant [7].

SSTs are usually unilateral except in rare cases. In the present case the tumor was observed to arise from the right ovary; 71% of SSTs have been found on the right side in the literature [2-4].

Macroscopically, they are greyish-white to yellow tumors with a smooth surface, varying in diameter from 2 cm to 17 cm at the widest point. Many are solid lesions containing small cysts and only two cases were reported as primarily cystic tumors [8, 9].

Microscopically, the SSTs are characterized by a pseudolobular pattern, in which cellular areas are separated by edematous and collagenous hypocellular tissue, pronounced sclerosis within the cellular nodules, a prominent vascularity and heterogeneity of the cell population [2-4].

Benign ovarian tumors associated with Meigs' syndrome are not common. Samanth and Black reported that 14.2% of ovarian benign stromal tumors were associated with ascites [10]. Young and Scully also pointed out that about 40% of fibromas over 10 cm in diameter are associated with ascites [11]. Ascites was observed only in one of the previously reported cases of ovarian sclerosing stromal tumors [12].

Pathologically, SST is occasionally confused with massive ovarian edema. Both lesions can occur in younger age groups. Preserved ovarian structures within the edematous stroma and absence of heterogeneity permit differentiation of massive ovarian edema from SST. The vacuolated cells in SSTs resemble signet ring cells in Krukenberg tumors, although these vacuolated cells do not contain PAS positive materials and present with benign appearing nuclei. Moreover, signet ring stromal tumor and ovarian myxoma should be distinguised.

All the SSTs encountered to date have been benign and recurrence has not been reported [1-12].

In summary, when assessing an ovarian tumor with ascites, a stromal tumor should be considered in the differential diagnosis. In addition, if it occurs in young women with metrorrhagia and infertility, then a sclerosing stromal tumor should be regarded as a strong possibility.

References

[1] Chalvardjian A., Scully R.E.: "Sclerosing stromal tumors of the ovary". *Cancer,* 1973, *31*, 664.

[2] Marelli G., Carinelli S., Mariani A., Frigerio L., Ferrari A.: "Sclerosing stromal tumors of the ovary: Report of eight cases and review of the literature". *Eur. J. Obstet. Gynecol. Reprod. Biol.,* 1998, *76,* 85.

[3] Saitoh A., Tsutsumi Y., Osamura Y., Watanabe K.: "Sclerosing stromal tumors of the ovary. Immunohistochemical and electron-microscopic demonstration of smooth-muscle differantiation". *Arch. Pathol. Lab. Med.,* 1989, *113,* 372.

[4] Stylianidou A., Varras M., Akrivis C., Fylaktidou A., Stefanaki S., Antoniou N.: "Sclerosing stromal tumors of the ovary: a case report and review of the literature". *Eur. J. Gynaecol. Oncol.,* 2001, *22,* 300.

[5] Shaw J.A., Dabbs D.J., Geisinger K.R.: "Sclerosing stromal tumors of the ovary: an ultrastructural and immunohistochemical analysis with histogenetic considerations". *Ultrastruct. Pathol.,* 1992, *16,* 363.

[6] Cashell A.W., Cohen M.L.: "Masculizing sclerosing stromal tumors of the ovary during pregnancy". *Gynecol. Oncol.*, 1991, *43,* 281.

[7] Yuen B.H., Robertson I., Clement P.B., Mincey E.K.: "Sclerosing stromal tumors of the ovary". *Obstet. Gynecol.*, 1982, *60*, 252.

[8] Gordon C.I., Slotnick E.: "Sclerosing stromal tumors of the ovary: report of a case". *J. Am. Osteopath. Assoc.*, 1983, *83,* 163.

[9] Hsu C., Ma L., Mak L.: "Sclerosing stromal tumors of the ovary". *Int. J. Gynecol. Pathol.*, 1983, *2,* 192.

[10] Samanth K.K., Black W.C.: "Benign ovarian stromal tumors associated with free peritoneal fluid". *Am. J. Obstet. Gynecol.*, 1970, *107*, 538.

[11] Young R.H., Scully R.E.: "Ovarian sex cord stromal tumors. Recent progress". *Int. J. Gynecol. Pathol.,* 1982, *1,* 101.

[12] Tsukamoto N., Nakamura M., Ishikawa H.: "Sclerosing stromal tumors of the ovary". *Gynecol. Oncol.,* 1976, *4*, 335.

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