BRIEF COMMUNICATIONS

ASCITES AND ELEVATED ANDROGEN LEVEL IN A PREGNANT PATIENT WITH AN OVARIAN SCLEROSING STROMAL TUMOR

Soon-Cen Huang,1 Hsiang-Chun Chen,1 Kong-Chao Chang,2 and Cheng-Yang Chou1

Abstract: We report the case of a 31-year-old woman with an androgen-producing sclerosing stromal tumor found during the eighth week of gestation in association with ascites and elevated serum androgen and cancer antigen (CA)-125 levels. The combined features of ascites, elevated serum androgen and elevated CA-125 in association with this rare type of ovarian tumor is unusual. Surgical removal of the tumor resulted in relief of symptoms, resolution of ascites, and decreases in serum androgen and CA-125 levels. The pregnancy was uneventful.

Key words: Sex cord-stromal tumor; Fibroma; Pregnancy; CA-125 antigen; Ascites


Sclerosing stromal tumor (SST), first described by Chalvardjian and Scully in 1973,1 is a rare type of ovarian tumor. To date, only 6 cases with SST diagnosed during pregnancy have been reported,2 and most of the cases were incidentally detected in the absence of specific symptoms. Here we report the case of a 31-year-old patient with such a tumor found during the eighth week of gestation. The tumor was complicated with ascites, an even rarer condition. The major presenting symptoms were progressive abdominal distention and a palpable pelvic mass. In addition, serum cancer antigen (CA)-125 and androgen levels were elevated.

Case Report

A 31-year-old pregnant woman, gravida 2 para 1, presented with progressive abdominal fullness and shortness of breath for 1 month. No pelvic tumor had been detected in previous pelvic sonography done 5 years ago. Review of her previous medical history and family history were not contributory except for deepening of her voice for several years. Physical examination revealed that the abdomen was distended and associated with shifting dullness, and a fist-sized movable mass was palpable at the right lower abdomen. No obvious signs suggestive of virilization such as acne or excessive hair growth were detected.

Laboratory examinations were all within normal reference limits except for an elevated serum CA-125 level of 396 U/mL (reference level < 33 U/mL). Ultrasound examination demonstrated an intrauterine pregnancy at 8 gestational weeks, a 7 x 6 x 6 cm right adnexal solid tumor, and profuse ascites (Fig. 1.) which extended upward to Morrison’s pouch of the abdomen.

Fig. 1. Pelvic sonography revealed an intrauterine fetus of 8 gestational weeks (S), a 7 x 6 x 6 cm solid mass (T) and ascites (A).
abdomen. Some intratumoral blood flow could be identified with Doppler flow mapping.

Due to progressive respiratory distress and suspicion of ovarian malignancy, explorative laparotomy was suggested and was accepted by the patient. At laparotomy, a right ovarian mass and approximately 1300 mL of ascites, straw yellow in color, were found. The uterus, left ovary, and tubes were unremarkable, and no evidence of intraperitoneal implants or enlarged lymph nodes was found. Right oophorectomy was performed and the specimen was submitted for pathologic evaluation.

Pathologic diagnosis by frozen section showed sclerosing stromal tumor of the right ovary. Examination of the right ovary revealed that the tumor weighed 200 g, and the cut surface was edematous, solid white and lobulated. Microscopic examination showed a pseudolobular pattern of cellular areas, separated by paucicellular edematous or myxomatous stroma as compared with normal ovarian tissue (Fig. 2A). The cellular areas were composed of an admixture of polygonal and spindle cells with vacuolated or eosinophilic cytoplasm (Fig. 2B). In some focal areas, dilated thin-walled blood vessels reminiscent of a hemangiopericytoma were observed. There were few mitotic figures. This characteristic appearance was diagnostic for sclerosing stromal tumor of the ovary and allowed differentiation from other tumors in the thecoma-fibroma group.

Biochemical analysis of ascites was transudative in character. Retrospective examination of preoperative serum androgen levels showed: testosterone, 19.9 ng/mL (reference level 0.1 to 1.2 ng/mL); 17-oxyprogesterone (17-OHP) 28.5 ng/mL (reference level 0 to 1.97 ng/mL); and dihydroepiandrosterone sulfate (DHEAS) 301 mg/mL (reference level 159 to 507 µg/mL), suggesting androgen secretion by this ovarian tumor.

The postoperative course was uneventful and the respiratory symptoms were relieved. Repeated sonographic examination 1 month after operation revealed a normal intrauterine pregnancy appropriate for gestational age and ascites was not detected. The serum androgen levels had returned to the normal reference range. The patient gave birth to a male baby at term without gross anomaly.

**Discussion**

Sclerosing stromal tumor is a rare type of ovarian tumor and has recently been described as a histologically and clinically distinct subgroup within the thecoma-fibroma spectrum of benign ovarian sex cord stromal tumors. Since the definition of this pathologic condition in 1973 by Chalvardjian and Scully, more than 80 cases have been reported. Most of the reported cases occurred in young women and only 6 cases have been reported during pregnancy. In most cases, the tumor was detected incidentally in asymptomatic patients. This tumor is rarely associated with endocrine presentations that have potential to produce steroid hormones, either androgens or estrogens. Only 2 cases of androgen-producing sclerosing stromal tumor during pregnancy have been reported. Both of them had signs of virilization, such as acne, excessive hair growth, and deepening of the voice. Deepening of the voice and elevated serum androgen levels were both observed in our patient, and decreased after surgical resection of the tumor. We therefore suspected that this androgen-producing tumor had existed in her body for years.

![Fig. 2. Microscopic findings of sclerosing stromal tumor. A) The pseudolobular pattern in this tumor was formed by cellular nodules separated by edematous connective tissue. B) The cellular pseudolobules were composed of collagen-producing spindle cells (arrowheads) and round vacuolated cells (arrows) [hematoxylin and eosin, x100].](image)
To our knowledge, ovarian sclerosing stromal tumors have not previously been reported in association with both spontaneous pregnancy and ascites. A woman with Gorlin’s syndrome was reported to have bilateral ovarian sclerosing stromal tumors in association with pregnancy following clomiphene therapy.\(^3\) Ascites was observed, but its relation to sclerosing stromal tumor, Gorlin’s syndrome-associated cancer or clomiphene therapy was not described. In addition, information regarding whether ascites transiently occurred or was persistent during pregnancy was lacking from the report.

Sclerosing stromal tumor of the ovary is characterized by a pseudolobular pattern of cellular areas separated by paucicellular edematous or sclerosing stroma. The cellular areas are composed of polygonal and spindle cells, which are surrounded by thin-walled blood vessels. Mitotic figures are distinctly uncommon. This above-mentioned appearance of sclerosing stromal tumor allows differentiation from other tumors in the thecoma-fibroma group.\(^1\) The edematous areas are localized only in the tumor part, allowing differentiation of massive edema of the ovary from other lesions. In massive edema of the ovary, normal ovarian tissues are dispersed by edematous stroma but no tumor tissue is found.\(^5\)

Because sclerosing stromal tumor is a subgroup of thecoma-fibroma, the mechanism of peritoneal effusion formation might be the same as that of fibroma. Fibromas are associated with ascites in approximately 10 to 15% of all cases. Though not well documented, the most likely pathogenesis ascribes the fluid formation to the filtration of interstitial fluid in the peritoneum through the tumor capsule, which does not have the capability of resorption.\(^6\) The hormonal milieu conferred by steroid production during pregnancy and possibly by sclerosing stromal tumor confirmed in some cases\(^7\) could additionally aggravate this transudative mechanism.

In addition to ovarian malignancy, CA-125 has been demonstrated to be elevated in many benign conditions. In this patient, the causes of CA-125 elevation could be due to the ovarian tumor, early pregnancy, ascites, or a combination of these conditions.

Due to the rarity of ovarian sclerosing stromal tumor and the unusual presentation such as progressive respiratory distress, as in our patient, it is not always possible to correctly predict its presence before surgery. Differentiation from other ovarian tumors or malignancy is even more difficult in pregnant women because tumor markers such as CA-125 may already be elevated and because some aggressive examinations may not be safe to the fetus. Careful preoperative evaluation and preparation, meticulous intraoperative management and regular postoperative follow-up are needed to ensure the well-being of both mother and fetus. Attention must also be given to the possible effects of hyperandrogenism on the fetus.

References