Sclerosing stromal cell tumor of the ovary in pregnancy: A case report

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Summary

A rare case of benign ovarian stromal cell tumor during pregnancy is presented. Because of a rapidly growing solid ovarian mass, 6 x 7 cm in diameter, a 21-year-old woman at 14 weeks of gestation was explored via laparotomy. Histopathological diagnosis was sclerosing stromal tumor of the ovary. She had no complaint of menstrual irregularities before pregnancy and there was no clinical or hormonal evidence of active androgenic hormone secretion. Immunohistochemical staining showed positive vimentin, smooth-muscle actin and desmin reactions. Sclerosing stromal tumor is a very rare condition in pregnancy and our case is only the eighth case detected during pregnancy according to the literature.

Key words: Sclerosing stromal tumor; Pregnancy; Ovary.

Introduction

Since the definition of sclerosing stromal cell tumor (SST) of the ovary in 1973 by Chalvardjian [1], over 80 cases have been reported. These tumors may be hormonally active or inactive. SSTs are benign neoplasms that usually occur in patients younger than 30 years of age [2]. Moreover, SST is a very rare entity in pregnancy and to date only seven cases have been reported [3, 4]. We present an additional case of SST of the ovary detected during pregnancy.

Case Report

A 21-year-old woman in her first pregnancy at a gestational age of 14 weeks was admitted to Zeynep Kamil Women and Children Diseases Education and Research Hospital on February 2001 because of a rapidly growing right adnexal tumor. The mass was detected on routine antenatal examination in the 11th week of the gestation. Ultrasonographic examination revealed a solid ovarian mass 4 x 3 cm in diameter. During follow-up three weeks later the mass measured 6 x 7 cm so the patient was admitted to the perinatology clinic. In her gynecological history she had been married ten months before and she had had regular menstrual cycles with 28-day intervals prior to pregnancy.

On pelvic examination, a 6 x 7 cm solid mass on the right side of the pelvis was palpated. Serum levels of total testosterone (40 ng/dl) and DHEA–S (90 mg/dl), free testosterone (1.1 ng/dl) were within normal limits. Tumor markers such as alfa-fetoprotein (0.13 ng/ml), carcinoembryonic antigen (0.8 ng/ml), cancer antigen (CA)-125 (24.45U/ml), and CA-19.9 (2.14U/ml) were also within normal limits. Human chorionic gonadotropin value was ignored because of pregnancy.

An immediate laparatomy was planned because of rapid growth of the mass. During explorative laparatomy, the uterus was in the midline with a gestational size of 14 weeks. The appearance of the left fallopian tube and ovary was normal. On the right, a solid rigid ovarian mass 6 x 7 cm in diameter with a regular surface and without any peripheral adhesions was detected. After excision of this mass, frozen section analysis revealed thecoma. Thus, a right salpingo-oophorectomy was performed.

The right ovary was 6 x 8.5 x 2.5 cm in size. On cross-sectioning, the ovarian tissue had a crescent shape and was 1.5 cm in its widest portion. The tumor measured 7.5 x 6 x 2 cm and was a pinkish-white colored solid mass with a well circumscribed pushing border with the residual ovarian parenchyme. No areas of cystic degeneration within the tumor were present.

On low-power microscopic examination a pseudolobulated pattern composed of both cellular and hypocellular areas was seen. In the hypocellular areas, edematous degeneration and collagenization existed and these areas were vaguely demarcated from the cellular areas. In the cellular areas, the cellularity was higher and nuclear enlargement more prominent. Small-to-medium sized blood vessels were very prominent particularly in the cellular areas and they showed a hemangiopericytomatous "staghorn" configuration in focal areas. These vessels, showing occasional dilatation and a sinusoidal appearance, were thin-walled and lined by a single layer of flattened endothelium (Figure 1). The cellular areas exhibited a heterologous cell population, luteinized theca cells with clear cytoplasm accompanied by rounded nuclei and scattered spindle shaped fibroblastic cells with eosinofilic cytoplasm accompanied by elongated nuclei (Figure 2). More than 60% of the cells of the tumor showed vimentin, smooth-muscle actin (SMA) and desmin positivity at immunohistochemical staining (BIOGEN-Neomarkers).

During follow-up the patient delivered a normal male infant at 38 weeks of gestation.

Discussion

Histopathological characteristics of the tumor were as follows [1]:

1) Pseudolobulation formed by cellular areas were separated by hypocellular collagenized areas.

2) Rich thin vascularity was seen in cellular areas.

Revised manuscript accepted for publication December 30, 2003

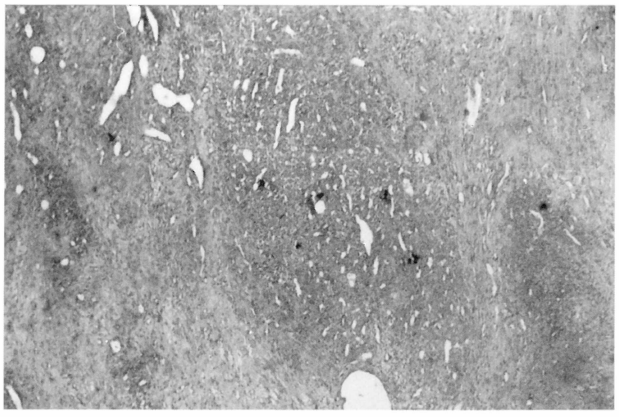

Figure 1. — Sclerosing stromal tumor-cellular islands are separated by paucicellular fibrous tissue creating a pseudolobular pattern. Notice the prominent vascular pattern which resembles that of a hemangiopericytoma (H & E x 50).

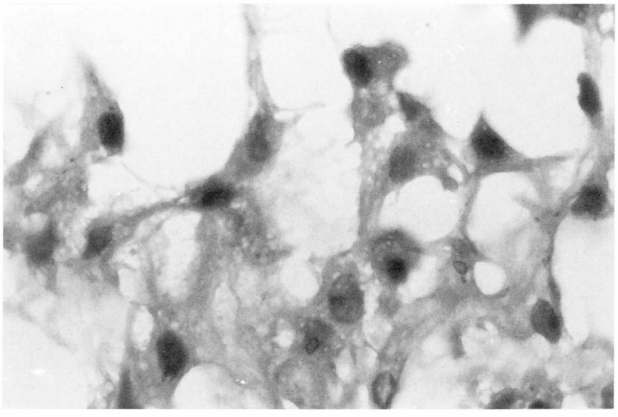

Figure 2. — Admixture of spindle cells and polygonal cells with vacuolated cytoplasm can easily be seen. An edematous area is evident at the top (H & E x 400).

3) Two types of cells were seen; theca-like cells containing lipid globules with cytoplasmic vacuolization and spindle-like cells forming collagen fibers.

4) Marked sclerosis was also detected outside the lobule.

The tumor cells were heterogeneous and exhibited a spectrum ranging from theca cells to ovarian stromal cells forming reticulin collagen fibers. In our case, smooth muscle-like cells were also present as demonstrated by immunohistochemical staining. This was a useful marker in distinguishing SST from thecomas and fibromas. Saitoh *et al.* [5] showed that cases of SST showed desmin (+), vimentin (+) and smooth-muscle actin (+). In the cellular areas in our case the cellularity was higher and nuclear enlargement was more prominent than in the cases in the literature. This was probably due to the hormonal effects of pregnancy. The number of luteinized theca-like cells was higher than in the reports of the previous literature [5], also probably due to the hormonal effects of the pregnancy. There are also some differences between SST and the thecoma-fibroma group regarding clinical presentation. The thecoma-fibroma group presents in the fifth and sixth decades of life [6]. The age of patients with SST are younger, usually under 30 years of age [2]. Our patient was 21 years old. SSTs have a prominent vascularity unlike thecomas and fibromas. Hyaline plaque, which is common in thecomas and fibromas, is not present in this group of tumors. Ascites is frequently encountered in the thecoma and fibroma group but rarely present in SST [4, 6]. No ascites was detected in our case. Cases of SST are reported to present menstrual irregularities and a history of infertility due to hormonal imbalance. In a review of the literature only seven cases in pregnancy have been reported up to date and three of these cases were with maternal virilization [3]. In the seventh case of the literature review, there was also an association with ascites, elevated serum androgen and CA-125 levels [4]. Our case is the eighth case of SST during pregnancy. Its solid and rapidly growing nature, as in our case, and its rarity strongly suggest malignancy and mandates an accurate histopathological diagnosis. Surgery with removal of the involved ovary is a sufficient treatment modality to cure these tumors. Prognosis is excellent.

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