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CASE REPORT

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Unclassified tumor of the gonadal stroma in a pregnant woman

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Abstract Gonadal sex cord stromal tumors contain some of the most morphologically interesting neoplasms of the gonads, and these lead to many important issues in differential diagnosis. Granulosa cell tumors are much more common in females. Unclassified sex cord stromal tumor is rare. We report a case of a 22-year-old woman with acute ruptured unclassified sex cord stromal tumor of the right ovary during pregnancy. The biological behavior of unclassified sex cord stromal tumors resembles that of Sertoli-Leydig cell tumors of intermediate differentiation rather than poorly differentiated tumors, which may have been expected in view of the lack of specific differentiation. This finding is important with regard to postoperative management.

Keywords Unclassified · Gonadal stromal tumor · Pregnancy

Introduction

Sex cord stromal ovarian tumors are rare malignancies, accounting for approximately 5% of all ovarian tumors [1]. Most sex cord stromal tumors of the ovaries can be composed of Leydig, Sertoli, or granulosa cell types existing as either a pure or mixed population. A minority of sex cord stromal tumors show no definitive differentiation and are classified as sex cord stromal tumors of the unclassified type or undifferentiated sex cord stromal tumors, comprising about 10% of all sex cord stromal tumors [2, 3]. Most classification difficulties occur with tumors that have features between those of granulosa and Sertoli-Leydig tumors, or with areas suggestive or diagnostic of both granulosa and Sertoli-Leydig differentiation [4]. The difficulty in classifying these tumors is their low degree of differentiation. They may be associated with poor prognosis [4]. We report an extremely rare tumor of the gonadal stroma during pregnancy, as well as present a review of the literature.

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Case report

A 22-year-old, gravida 1, para 0 woman presented to our emergency department with complaints of diffuse abdominal pain and intractable nausea and vomiting for 3 days' duration at 32 weeks' gestation. The patient' abdomen was distended, and on obstetric examination the cervix was 5 cm dilated and about 80% effaced, with intact membranes. An ultrasound scan revealed ascites with hyperechogenic particles, an appropriately sized fetus, and a right complex adnexal mass about 15×15 cm in size and an appropriately sized fetus. The CA125 level was elevated to 650 U/ml.

The patient underwent an emergency exploratory laparotomy. Intraoperative findings included approximately 1 l of serohemorrhagic fluid and a solid, ruptured, 15×15-cm right ovary containing lipoid areas. The left ovary and fallopian tube appeared normal. The decision was made to perform a conservative procedure to preserve future fertility; therefore, a cesarean section and a right salpingo-oophorectomy were done. The male infant weighed 1,600 g with Apgars of 7 at 1 min and 9 at 5 min. The patient's postoperative course was uneventful, and she was discharged home on the 7th postoperative day.

On pathologic examination, the tumor was 15×14×4 cm in diameter, and the surface was smooth. In cut section, predominantly solid areas revealed gray-white color and firm consistency. In addition, cystic areas were filled with fluid, and there were some areas of hemorrhage and necrosis attracted attention. On microscopic examination, the tumor was composed of spindle and polygonal cells.

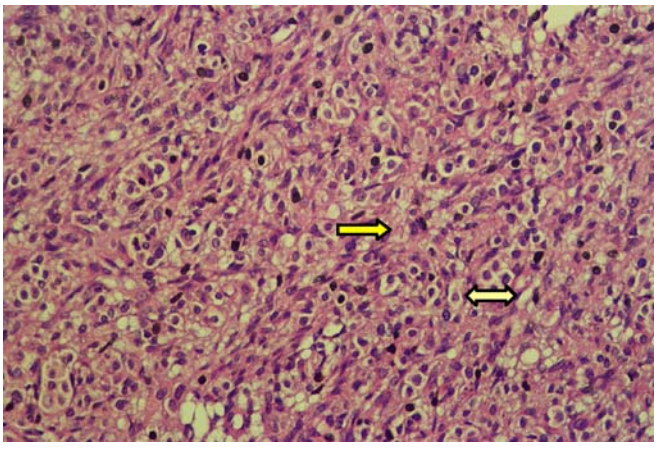

Fig. 1 Spindle cells (*arrow*) and polygonal cells in nest (*double-headed arrow*)

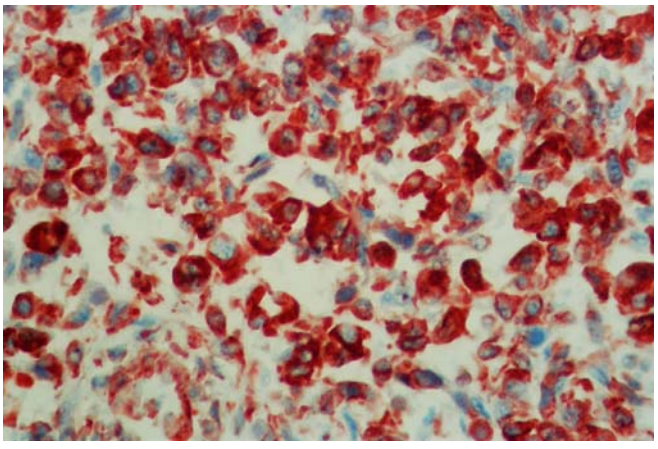

Fig. 3 Vimentin staining in tumor cells (×40)

The latter were large and pale eosinophilic cells in a nestlike pattern with ovoid, indented nuclei (Fig. 1). The nest of polygonal cells was surrounded by reticulum fiber (Fig. 2). On immunohistochemical examination the tumor cells were stained with vimentin (Fig. 3), and only polygonal cells were stained with inhibin (Fig. 4).

The patient's case was reviewed in the weekly tumor board, and final pathology was consistent with stage IIIC unclassified sex cord stromal tumor. The patient received four cycles of chemotherapy with bleomycin, etoposide, and cisplatin (BEP regimen). She has remained disease-free as of 10 months postoperatively, and all tumor markers have normalized.

Discussion

The number of adnexal masses diagnosed concurrently with pregnancy has increased due to routine use of ultrasonography during pregnancy. Adnexal masses have been noted to occur in up to 1% of all gestations [5]. Gross and microscopic examination to exclude a malignant neoplasm is an indication for operative intervention. The reported incidence of ovarian cancer in pregnancy ranges from 1:5,000 to 1:47,000 live births, with 2–6% of persistent adnexal masses being malignant [6, 7]. Epithelial cancers and tumors with low malignant potential are the most common ovarian cancers found in pregnancy [5]. In a study of 36 cases of gonadal stromal tumors during pregnancy, there were only six cases of unclassified sex cord stromal tumors [8].

In 1976 Fox and Langley noted that "tumors formed of immature granulosa-theca cells resemble closely those formed of immature Sertoli-Leydig cells, often to the extent that it is quite impossible to differentiate the two" [9]. Young and Scully have stated that "it is to some extent a matter of personal preference whether one gives more weight to the Sertoli cell or granulosa-theca patterns of such tumors and accordingly places them in the Sertoli-Leydig cell or granulosa cell tumor category" [2]. It is clear that some gonadal stromal tumors defy current classification schemes. It has been suggested that the paucity of reported cases of unclassified sex cord stromal tumors is due to hesitation by pathologists to report cases without definite diagnoses as well as to the inherent subjectivity in making this diagnosis [4, 10]. Young and Scully currently include these tumors in the unclassified category [2].

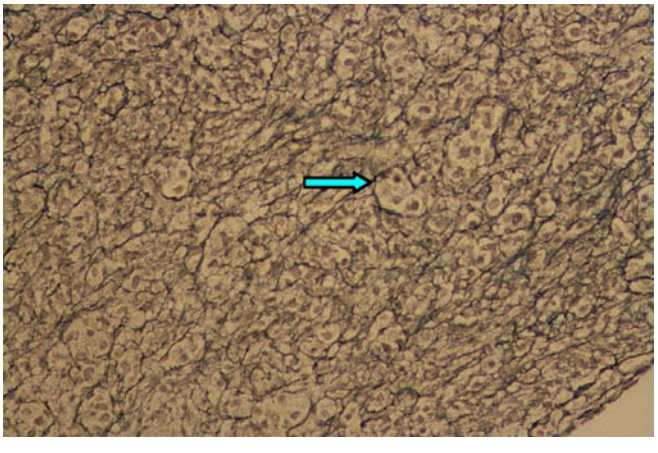

Fig. 2 The nest of polygonal cells was surrounded by reticulum fiber (*arrow*). (Reticulum staining ×20)

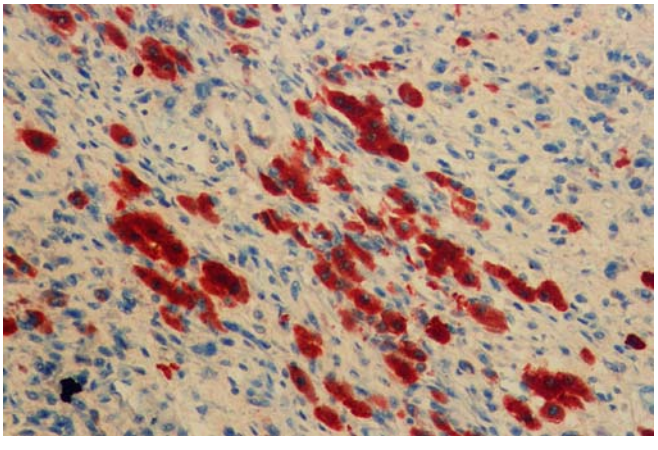

Fig. 4 Inhibin staining in nest of the polygonal cells (×40)

Some gonadal stromal tumors, particularly those with features between those of Sertoli-Leydig and granulosa tumors, are difficult to classify [2]. In most of these cases, the differential diagnosis is between granulosa and Sertoli-Leydig types. The behavior of this group of tumors has not been adequately studied. Prognostic factors associated with sex cord stromal tumors include age, stage of disease, tumor size, extent of surgery, and other histologic parameters [11]. The prognosis for patients with unclassified gonadal stromal tumors is similar to that for patients with granulosa or Sertoli-Leydig cell tumor [4]. Tumor size has been recognized as an important prognostic factor in many reports; previous studies found that woman with larger tumors (≥10 cm) were more likely to have recurrence [11, 12]. Another very significant prognostic factor is tumor stage at presentation.

We performed conservative surgery for our patient because no significant difference in overall survival exists between unstaged and staged patients [11]. In one study, pregnant patients, with one exception, were initially treated by conservative surgical procedures. Hysterectomy and salpingo-oophorectomy were performed at a second operation; no residual tumor was found in any of the specimens [8]. In our case, the tumor size was approximately 14 cm, and the stage was IIIC. Because of risk of recurrence, the patient received four courses of the BEP regimen. She has remained disease-free 10 months postoperatively, and all tumor markers have normalized.

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