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Ovarian steroid cell tumor, not otherwise specified: A case report and literature review

LILI QIAN, ZHEN SHEN, XUEFEN ZHANG, DABAO WU and YING ZHOU

Department of Obstetrics and Gynecology,
Anhui Provincial Hospital Affiliated to Anhui Medical University, Hefei, Anhui 230001, P.R. China

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Abstract. Steroid cell tumors (SCT), not otherwise specified (NOS) are particularly rare ovarian sex cord-stromal tumors, which comprise <0.1% of all ovarian tumors. These tumors are uncommon in patients' prior to puberty without any typical syndromes involving hirsutism, virilization and hypertension. We here in present the case of a 5-year-old female patient who presented with sudden abdominal pain, repeated vomiting and a pelvic mass. Our patient underwent urgent exploratory laparotomy and right salpingo-oophorectomy and the histopathological examination revealed an ovarian SCT-NOS. The patient has been followed up for 5 years since the surgery, without evidence of disease recurrence. The purpose of this study was to discuss the available information on the presentation, diagnosis and recommended treatment of ovarian SCT-NOS; and describes the immunohistochemical characteristics of these tumors.

Introduction

Ovarian steroid cell tumors (SCT) are rare sex cord-stromal tumors of the ovary and comprise <0.1% of all ovarian tumors (1). Based on the cell of origin, they may be divided into three subtypes: Leydig cell tumors arising from Leydig cells in the hilum of the ovary, stromal luteomas arising from ovarian stromal cells, and steroid cell tumors not otherwise specified (NOS). The last subtype makes up ~2/3 of SCTs and tends to affect younger women (mean age, 43 years). SCT-NOS are usually benign; however, clinically malignant behavior, such as peritoneal metastases, occurs in 25-40% of the cases (2,3). We herein report a case of an ovarian SCT-NOS in a 5-year-old female Chinese patient who presented with sudden abdominal pain and repeated vomiting. Aspects of the presentation, diagnosis and treatment of these tumors are also discussed.

Correspondence to: Dr Ying Zhou, Department of Obstetrics and Gynecology, Anhui Provincial Hospital Affiliated to Anhui Medical University, Hefei, Anhui 230001, P.R. China
E-mail: caddie1234@gmail.com

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Case report

A 5-year-old female Chinese patient presented to the Department of Obstetrics and Genecology of the Anhui Provincial Hospital (Hefei, China) on 2/12/2011 with sudden abdominal pain and repeated vomiting. A computed tomography (CT) pelvic scan and ultrasound examination in another hospital on the previous day revealed a pelvic mass. The B-ultrasound scan in our hospital revealed a solid, right ovarian tumor sized 85x45x73 mm. On laboratory analysis, the serum carbohydrate antigen (CA) 19-9 level was 30.25 U/ml (normal, 0-34 U/ml); the β-human chorionic gonadotropin level was <0.10 IU/l (non-pregnancy, <0.5-2.9 IU/l); the α-fetoprotein level was 1.15 ng/ml (normal, 0-8 ng/ml); and the CA125 level was 37.71 U/ml (normal, 0-39 U/ml). The patient underwent urgent exploratory laparotomy, and the right ovary, sized 8x4x7 cm, exhibited a smooth, unruptured surface and was reversed 720 together with the right Fallopian tube and the right ovarian intrinsic ligament. The patient was then treated by right salpingo-oophorectomy. The histopathological examination revealed an ovarian SCT-NOS (Fig. 1), composed of cells positive for inhibin, vimentin, progesterone receptor, calretinin, somatostatin, synaptophysin, CD99, and weakly for D2-40, and negative for CD117, estrogen receptor, carcinoembryonic antigen, cytokeratin, human melanoma black 45, Melan A, S-100 and placental alkaline phosphatase (Fig. 2). The patient has been followed up for 5 years since the surgery, without evidence of recurrence or metastasis, or abnormalities on the laboratory test results.

Discussion

The term ‘steroid cell tumors not otherwise specified’ was first used by Scully and signifies that the cell lineage is not defined; thus, they cannot be categorized as either stromal luteomas or Leydig cell tumors (4,5). Approximately 56-77% of the cases are clinically associated with androgenic changes, such as hirsutism and virilization; 6-7% of the cases are clinically associated with Cushing's syndrome; and 25% of SCT-NOS are non-functional. Ovarian SCT-NOS may occur at any age (mean age, 43 years) and, occasionally, before puberty (6). In our case, the patient was aged 5 years, without any changes in blood pressure, virilization or hirsutism. The patient has been followed up for 5 years since the surgery, without disease

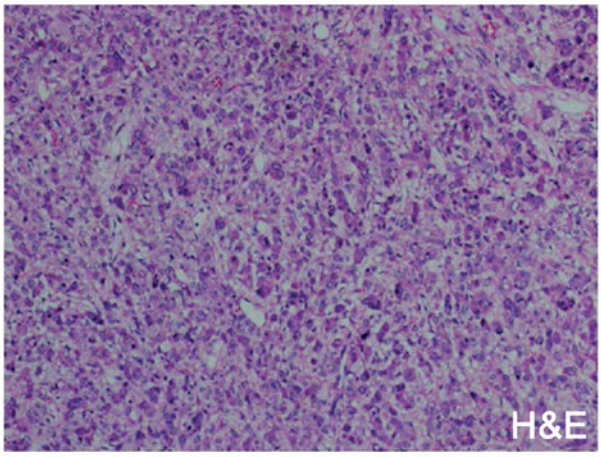

Figure 1. Hematoxylin and eosin (HE) staining of the ovarian tumor, not otherwise specified. The tumor was composed of cells with abundant eosinophilic to clear cytoplasm and round nuclei with prominent nucleoli (magnification, x160).

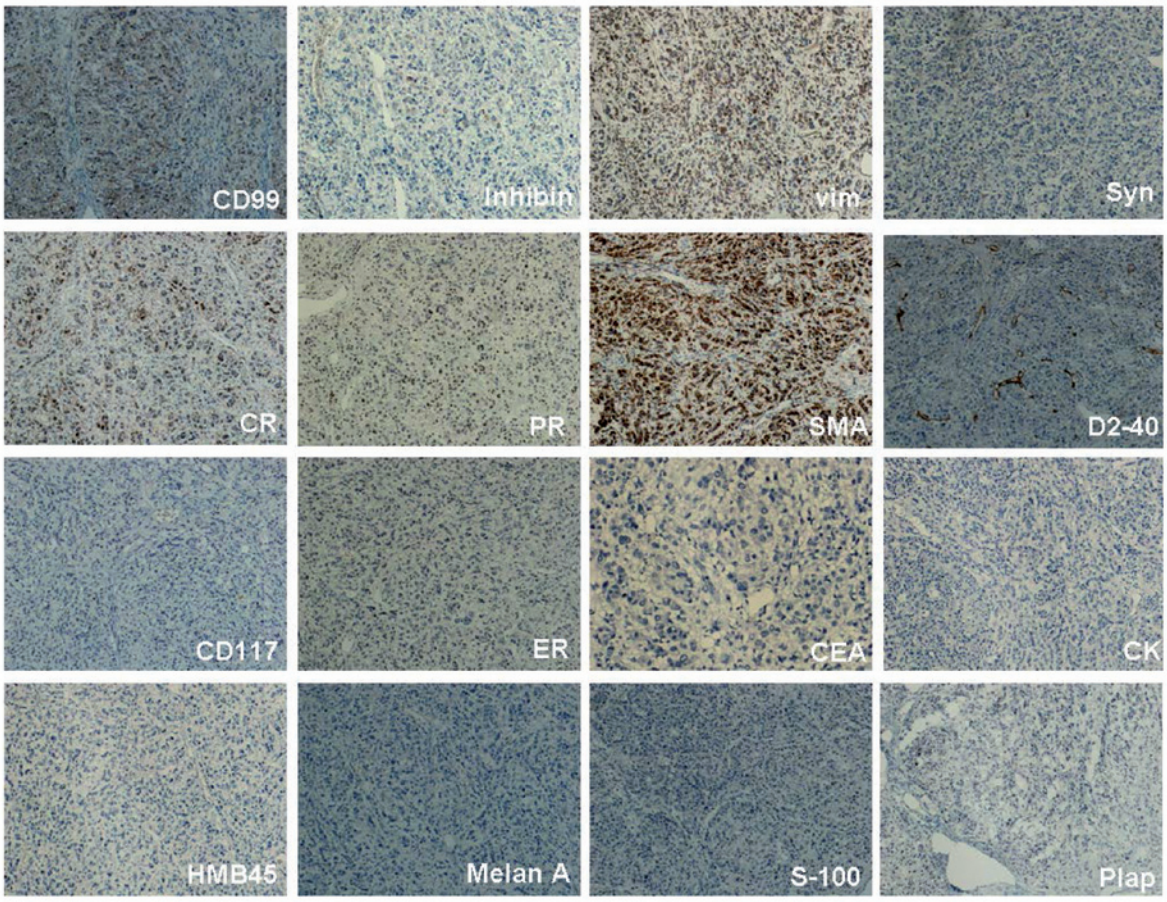

Figure 2. Immunohistochemical examination of the ovarian tumor (magnification, x160). The tumor cells were positive (brown staining) for inhibin, vimentin (vim), progesterone receptor (PR), calretinin (CR), somatostatin (SMA), synaptophysin (Syn), CD99 and weakly positive for D2-40, ER, estrogen receptor; CEA, carcinoembryonic antigen; CK, cytokeratin; HMB45, human melanoma black 45; Plap, placental alkaline phosphatase.

recurrence or metastasis. To the best of our knowledge, the youngest reported patient was aged 2.5 years when she first presented with virilization; the androgenic changes subsided following surgical removal of the enlarged cystic right ovary, but the patient succumbed to diffuse metastatic disease originating in an apparently unrelated undifferentiated sarcoma of the oropharynx 17 months after initial diagnosis (7).

According to the study of Hayes and Scully, the following pathological characteristics may indicate malignancy: ≥2 mitotic figures per 10 high-power fields is associated with a 92% risk of malignancy; necrosis with an 86% risk of malignancy; a diameter of >7 cm with a 78% risk of malignancy; hemorrhage with a 77% risk of malignancy; and grade 2 or 3 nuclear atypia with a 64% risk of malignancy (6). To date,

24 cases of ovarian SCT in young female patients aged 2.5-13 years have been reported, but none have been malignant (6,8-11). Therefore, ovarian masses suspicious for SCT in children at an early stage should be approached conservatively, unless distinct signs of metastasis are present at the time of surgery (12). Immunohistochemistry is particularly useful for the accurate diagnosis of SCTs. Moreover, unilateral salpingo-oophorectomy is generally considered to be adequate for pediatric patients with stage I a disease (6). In our patient, although the diameter of the mass was >7 cm, we opted for right salpingo-oophorectomy as the tumor was stage Ia. It is also necessary to monitor the patient's hormone levels as part of their postoperative follow-up (12).

To date, very few reports have investigated the efficacy of radiation or chemotherapy for SCTs (13). However, malignant SCT-NOS should be managed with total abdominal hysterectomy, bilateral salpingo-oophorectomy and sampling of pelvic and mesenteric lymph nodes and omentum followed by combination chemotherapy. Though a definitive chemotherapy regimen is not yet defined, Bleomycin, Etoposide and Cisplatin (BEP) is favored and often used. Chemotherapy maybe applied using the BEP regimen [bleomycin (20 U/m^2 every 3 weeks for 4 cycles), etoposide (75 mg/m^2 on days 1-5, every 3 weeks for 4 cycles) and cisplatin (20 mg/m^2 on days 1-5, every 3 weeks for 4 cycles)] (14), or other medications (6). Gonadotropin releasing hormone agonist has been used as therapy for recurrent malignant disease for its suppressive effect on ovarian steroidogenesis (15).

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