Case Report

Steroid Cell Tumor of the Ovary Presenting with Ascites: A Rare Neoplasm in a Postmenopausal Woman

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Abstract

Steroid cell tumors of the ovary are rare sex-cord stromal tumors, accounting for approximately 0.1% of all ovarian neoplasms. Majority of these tumors are benign, occur in pre-menopausal women and are associated with hyperandrogenism. However, around one-third of cases are malignant and do not present with hormonal manifestations. A 48-year-old post-menopausal woman presented with complaints of gradually increasing progressive abdominal distension over the past 3 months. She had a history of weight gain but denied any symptoms of virilization. On examination, abdominal distension associated with ascites was noted. Serum CA125 level was raised. Contrast-enhanced computed tomography revealed a solid right adnexal mass. Based on the clinical impression of epithelial ovarian malignancy, the patient underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy. Histopathological examination revealed steroid cell tumor of the not otherwise specified type in the right ovary with the capsular breach. However, all other organs, including the omentum were free of tumor. The index case is unique for its presentation in a post-menopausal woman, association with ascites, elevated CA125 levels and lack of any virilization manifestations. Establishing an early and accurate tissue diagnosis is essential so that appropriate surgical management can be done to prevent the development of metastases in potentially malignant cases.

Keywords: *Histopathology, immunohistochemistry, ovarian tumor, sex-cord stromal tumors, steroid cell tumor, virilization*

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Introduction

Steroid cell tumors of the ovary are rare sex-cord stromal tumors (SCST), accounting for approximately 0.1% of all ovarian neoplasms. These include three subtypes: stromal leuteomas, Leydig cell tumors, and steroid cell tumors, not otherwise specified (SCT-NOS).[1] Steroid cell tumor, NOS type is the more common, accounting for around 50%–60% of cases. As the name suggests, these tumors produce steroids and hence are associated with hyperandrogenism, leading to symptoms such as hair loss, hirsutism, temporal balding, and menstrual irregularities due to virilizing properties of the secreted hormones.[2] Owing to the hormonal symptoms, these patients are often diagnosed at an early stage, compared to ovarian epithelial neoplasms. However, around 20%–25% of patients lack these endocrine manifestations and hence may remain undiagnosed clinically for long periods.[3]

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Case Report

A 48-year-old post menopausal woman, presented with a history of progressively increasing abdominal distension and weight gain over the past 3 months. There was no associated abdominal pain, alteration of bowel/bladder habits, or vaginal discharge/bleeding. Her menstrual and obstetric history was not significant. She denied the intake of hormones or drugs and had not undergone any surgical intervention in the past.

On examination, her abdomen was distended with the presence of ascites. She did not have increased facial hair or acne. Contrast-enhanced computed tomography revealed a solid right adnexal mass measuring 8.9 cm × 11.5 cm × 9.5 cm. CA125 levels were raised (472

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U/mL), however, other tumor markers (carcinoembyonic antigen, alpha fetoprotein, lactate dehydrogenase and CA 19-9) were within the normal limits. Based on the clinical, biochemical and radiological findings, a provisional diagnosis of an epithelial ovarian malignancy was made.

The patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy. Intra-operatively, 500 ml of straw colored fluid was found in the peritoneal cavity; however, no tumor deposits were noted.

Grossly, the right ovarian mass measured 11 cm × 9.5 cm × 8 cm with the capsular breach. On cut-section, the tumor was relatively well-circumscribed, predominantly solid (98%) with occasional small cystic areas filled with thick-mucoidy secretions (2%). The solid areas had a lobulated appearance, were firm and yellowish [Figure 1]. Left ovary measured 2.3 cm × 1.8 cm × 1 cm, bilateral fallopian tubes measured 5cm each, uterus measured 6.5 cm × 6 cm × 5 cm, and were grossly within the normal limits. Microscopically, the tumor showed a lobulated appearance with tumor cells arranged predominantly in diffuse sheets. The lobules were separated by thin fibrovascular septae. The tumor cells depicted mild pleomorphism, were round to polygonal with distinct cell membranes, round-to-oval centrally placed nuclei, vesicular chromatin, prominent nucleoli, and abundant clear to vacuolated cytoplasm. The nuclear atypia was mild, being grade 2. The mitotic index was <2/10 high power fields. Tumor necrosis or Reinke crystals were not seen. The capsular breach was confirmed microscopically. Based on the histopathologic features, a diagnosis of steroid cell tumor, NOS was rendered. All other organs, including the omentum, were free of tumor. Immunohistochemistry for inhibin showed granular cytoplasmic positivity in the tumor cells [Figure 1].

Discussion

SCSTs of the ovary are rare and account for 4%–6% of all ovarian tumors.[4] Ovarian steroid cell tumors (OSCTs) are much rarer, with a reported frequency of around 0.1% of all the ovarian tumors.[5] Most commonly, OSCT is seen in premenopausal women with a mean age of 43 years.[6] The most common clinical presentation is abdominal pain and virilizing symptoms.[7] Presentation in postmenopausal women is rare and establishing an early clinical diagnosis in such cases becomes challenging, especially in the absence of virilizing manifestations.

OSCTs are usually benign, solid, and unilateral.[8] Approximately 25% of cases of SCT-NOS type are asymptomatic or present without hormonal manifestations. In addition, malignant OSCTs are also hormonally inactive.[3] The presence of ascites has been rarely noted in these tumors.[8] The index case presented with ascites without any symptoms of virilization. In addition, she had elevated CA125 levels, which also is an infrequent finding in OSCTs.[8] The most plausible explanation for this could be the mechanical irritation of the mesothelium by associated ascites.

A definite diagnosis can be established only by histopathological examination. Microscopically, the tumor cells are arranged diffusely in sheets, are polygonal, having round central nuclei with vesicular chromatin, prominent nucleoli, and abundant pale to eosinophilic and vacuolated cytoplasm. Lipid stains like oil red-O highlight the cytoplasmic lipids droplets. On immunohistochemistry, these tumor cells are positive for sex-cord stromal markers like inhibin and calretinin. The differential diagnoses include thecoma, clear cell carcinoma, metastatic renal cell carcinoma and rarely pheochromocytoma. The salient features that can help in differentiating these histopathologic mimics are listed in Table 1.

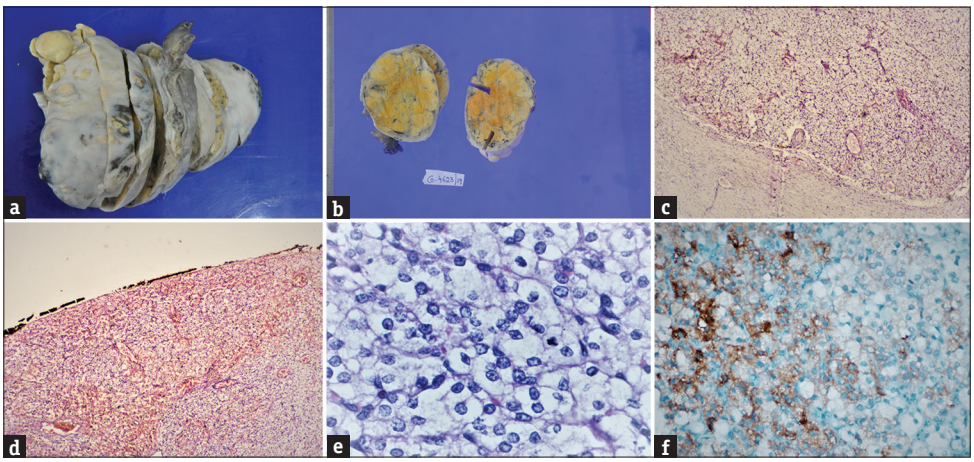

Figure 1: (a) Outer surface of the right ovarian mass showing lobulated appearance and capsular breach by the tumor; (b) Cut section of the ovarian mass showing solid, yellowish, lobulated tumor; (c) Relatively circumscribed tumor with tumor cells arranged in sheets (H and E, ×10); (d) Section showing capsular breach by tumor cells (H and E, ×4); (e) Section showing sheets of polygonal tumor cells with well-defined cell membranes, central nuclei with prominent nucleoli and abundant amount of clear to vacuolated cytoplasm (H and E, ×40); (f) Immunohistochemistry for Inhibin showing granular cytoplasmic positivity in the tumor cells (Inhibin, ×20)

Table 1: Histopathologic differential diagnoses of steroid cell tumor

Tumor	Age	Symptoms	Gross	Microscopy	IHC
Thecoma	Postmenopausal	Abnormal uterine bleeding, endometrial carcinoma	Small, solid, yellowish	Polygonal to round cells with abundant eosinophilic to vacuolated cytoplasm, hyaline plaques, abundant reticulum	Inhibin
Leydig cell tumor	Postmenopausal	Virilization, abdominal mass	Solid, yellowish	Polygonal cells with abundant dense eosinophilic granular cytoplasm with Reinke crystals	Inhibin
Clear cell carcinoma	Postmenopausal	Pelvic endometriosis, paraneoplastic hypercalcemia	Solid-cystic	Tubulocystic, papillary pattern	Cytokeratin, HNF1β
Metastatic RCC	Elderly	Weight loss, history of renal mass/hematuria	Solid, or variegated appearance with hemorrhage, necrosis	Sheets, papillae of cells with abundant clear cytoplasm with pleomorphic nuclei	CD10, Dual positivity for cytokeratin and vimentin
Pheochromocytoma	Young adults	Hypertension weight loss	Solid yellowish	Polygonal cells in nests with abundant cytoplasm with fine eosinophilic granules, Zellballen pattern	Synaptophysin ChromograninS-100 in sustentacular cells

IHC: Immunohistochemistry, HNF1β: Hepatocyte nuclear factor 1β, RCC: Renal cell carcinoma

Although majority are benign, around one-third can be clinically malignant.[9] Associated ascites, weight loss, and anorexia favor malignant behavior of the tumor. The index patient was post-menopausal and presented with abdominal distension without any hormonal symptoms. She had ascites, which favors malignant nature. The most predictive malignant features of OSCTs, as reported in the literature, are the presence of two or more mitotic figures per 10 high-power fields; necrosis; diameter of 7 cm or greater; hemorrhage; and grade 2 or 3 nuclear atypia.[10] In the index case, the tumor diameter was >7 cm and the nuclear atypia was grade 2. In addition, there was capsular breach; however, no peritoneal or pelvic tumor deposits.

Conclusions

OSCT is an extremely rare neoplasm. Although majority are benign, one-third can be malignant. The index case is unique for its presentation in a post-menopausal woman, association with ascites, elevated CA125 levels and lack of any virilization manifestations. Establishing an early and accurate tissue diagnosis is important for prompt surgical management to prevent the development of metastases in such cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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