Case Report

Sclerosing Stromal Tumor of the Ovary: A Successful Laparoscopic Approach

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Abstract

Sclerosing stromal tumors (SSTs) are a rare type of benign tumors of the ovary, representing 6% of sex cord tumors subtype. We report a case of SSTs affecting a young female patient presenting with abdominal pain and a pelvic mass on imaging examination. The patient underwent laparoscopic right salpingo-oophorectomy, and the pathology report confirmed the diagnosis of SSTs. A review of the literature with the typical pathological and imaging features of SSTs as well their management is performed.

Keywords: Ovarian neoplasms, sex cord-gonadal stromal tumors/pathology, sex cord-gonadal stromal tumors/surgery

Introduction

Sclerosing stromal tumors of the ovary (SSTs) are an extremely rare benign tumor of ovarian theca cells.[1]

Representing 6% of sex cord stromal tumors of the ovary, they were first described in 1973 by Chalvardjian and Scully with 301 cases reported to date.[2,3]

SSTs usually affects young patients, in their second or third decades of life and clinical presentation include menstrual irregularities, pelvic pain, and signs and symptoms related to an ovarian mass, most frequently unilateral.[4] Nevertheless, etiology is still unknown.[5]

Differential diagnosis with other ovarian tumors, including the malignant ones, can be challenging.[1] In most cases, the diagnosis is made by the pathologic examination during or after surgery.[6]

Since this rare benign tumor of the ovary often affects the woman of child-bearing age, additional efforts toward a conservative approach and fertility spearing surgery should be offered. Although of the 301 cases described in the literature, only a few reports a laparoscopic approach of the tumor.[1,2,6-8]

Laparoscopic surgery is possible, desirable, and should be offered to these young patients like the present case report.

Case Report

An 18-year-old female patient was referred to our emergency department for abdominal pain and a pelvic mass.

The patient was a healthy nulliparous woman without medical or surgical history of concern, neither familiar history noteworthy. She attended the medical emergency department for abdominal pain localized in the right iliac fossa and hypogastric region with months of evolution. The patient denied other symptoms such as menstrual irregularities, vomiting, nausea, anorexia, fever, or other complaints.

On physical examination, a large abdominopelvic mass was palpable on the right iliac fossa, with no signs of peritoneal

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irritation. Since the patient was a virgin, the gynecological examination was not performed.

Laboratory results were unremarkable, including negative tumor markers.

An ultrasound of lower abdomen and the pelvis was requested and confirmed a 9 cm mass localized on the right adnexal topography. The mass had cystic and solid components and was vascularized, with no apparent signs of adnexal torsion.

The abdomino-pelvic computed tomography scan and the magnetic resonance imaging (MRI) [Figure 1] described a polylobulated pelvic mass originated in the right adnexal area, measuring 7.7 cm × 6.9 cm × 9.8 cm, highly vascularized, with several calcifications and a hypocaptating central zone. There was also a small amount of ascitic fluid, but there were no signs of adenopathies or alterations on the other structures.

A diagnostic laparoscopy was proposed and accepted by the patient. After the introduction of the laparoscope, a solid lobulated mass with 10 cm of the right ovary was confirmed. There was torsion of the referred mass without vascular compromise.

Ascites was also confirmed and aspirated (about 200 mL). There were no other macroscopic alterations seen in the abdomino-pelvic cavity.

A right salpingo-oophorectomy was performed, and the suprapubic trocar incision was extended to allow the extraction of the tumor in the endobag.

Intraoperative frozen section examination [Figure 2] disclosed a benign neoplasm composed of spindle-shaped cells with a morphology compatible with sex cord-stromal tumor.

Macroscopic examination [Figure 2a] showed a multinodular mass of the ovary measuring 10 cm × 8 cm × 6.5 cm with a whitish and vascularized capsule. The cut surface revealed a lobulated solid mass, with a central scar, and areas of cystic degeneration.

Microscopically [Figure 2b and c], the neoplasm had cellular lobules in a background of hypocellular collagenous or edematous stroma, branching and thin-walled vessels, and two types of cells, bland spindle eosinophilic cells, and epithelioid cells with abundant clear cytoplasm.

The immunohistochemical study showed positivity for inhibin and alpha smooth muscle actin (SMA) and negativity for AE1AE3.

The final diagnosis was SST of the ovary.

Currently, the patient is under follow-up (1 year and half) with no signs of recurrence.

Discussion

SSTs are a rare type of sex cord tumor. These tumors are divided in granulosa stroma cell tumors, Sertoli's stromal tumors, steroid cell tumors, and other types. The first group is subdivided in granulosa cell tumors, thecomas, fibromas, and SSTs.[4]

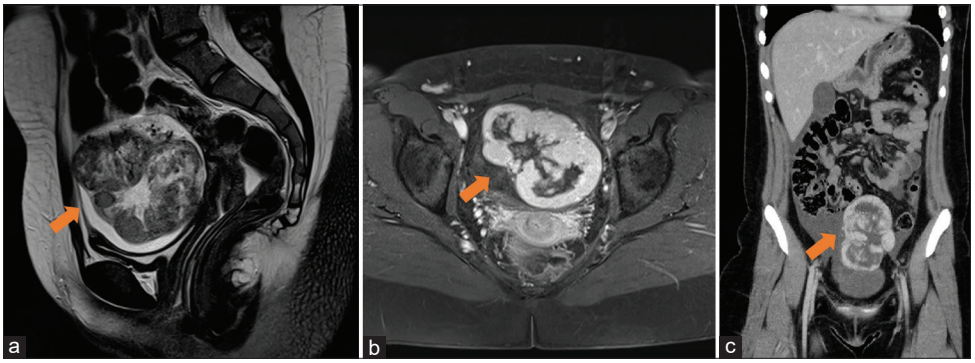

Figure 1: T2-weighted magnetic resonance imaging revealing an ovoid solid tumor of the ovary (arrow) on sagittal section (a), axial section (b), and coronal section (c)

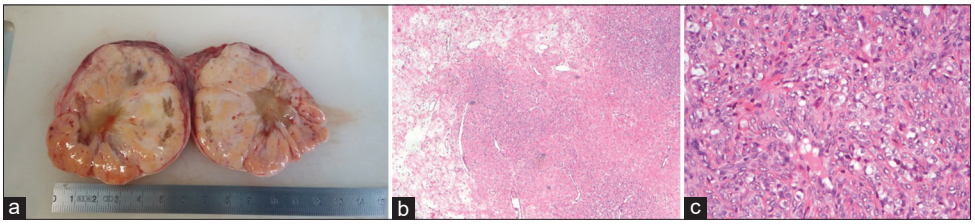

Figure 2: (a) Macroscopic analysis of 6 cm ovarian tumor (lobulated surface and a central scar). (b) Microscopy examination: Cellular and hypocellular areas (edema or sclerosis) and a vascularized stroma. (c) High magnification showing two types of cells (spindle eosinophilic cells and epithelioid cells)

The etiology of SSTs is still unknown, although some authors defend that these tumors arise from pluripotent immature stromal cells of the ovarian cortex.[2,9]

SSTs represent about 6% of ovarian sex cord tumors and usually affect patients during their second or third decade of life.[2,4] Common clinical presentation includes abdominal pain and a palpable abdomino-pelvic mass, as well as menstrual complaints.[10]

They also may present signs or symptoms related with estrogenic or androgenic augmented activity, although most tumors are hormonally inactive. Ascites is rare.[4]

The imaging features on ultrasound include a unilateral large mass on the adnexal region with a solid predominant component with hypoechoic areas. Intense peripheral vascularization is also common. On MRI, a large solid heterogeneous mass with high intensity sign in the cystic areas is usually reported.[4]

SSTs can be easily misdiagnosed as malignant tumors of the ovary.

Differential diagnosis should include other sex cord tumors such as fibroma and thecoma (although these tumors frequently occur in the fifth and sixth decades of life) but also ovarian malignant tumors.[2]

A pseudolobular architecture, a collagenous or edematous stroma in hypocellular areas, two types of cells without atypia, and positivity for inhibin and other sex cord markers are characteristic of these tumors, ruling out other sex cord tumors like fibroma and thecoma. Immunohistochemically, the tumor cells are positive for vimentin, inhibin, SMA, and alpha inhibin and negative for epithelial markers.[2,11]

A conservative surgical approach should be offered, since recurrence has not been reported yet.[1,2,8]

Conclusion

SSTs of the ovary should be considered in the differential diagnosis of unilateral heterogeneous ovarian masses in young female patients. As so, clinical presentation and typical imaging features must be known.

A fertile sparing surgery with a minimally invasive approach should be performed because the published studies agree in the benign nature of these neoplasms, and there is no case of recurrence reported.

Ethical approval

This study was approved by the Institutional Ethical Committee of Centro Hospitalar Vila Nova de Gaia/Espinho (IRB number 38/2020 obtained on January 23, 2020).

Declaration of patient consent

The authors certify that they have obtained an appropriate patient consent approved by the hospital ethical council. In the form, the patient has provided consent for images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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