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A Sertoli-Leydig ovarian tumor presenting as ovarian torsion: A case report

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ABSTRACT

Torsion occurs as a complication in 10% of cases of ovarian tumors. It predominantly occurs in benign ones, while malignant tumors are less prone to torsion. Sertoli-Leydig cell tumors are highly unusual sex cord-stromal tumors of the ovary, accounting for less than 0.2% of all ovarian cancers.

A 39-year-old patient presented to the emergency department with a Sertoli-Leydig cell tumor diagnosed due to ovarian torsion. The clinical presentation was characterized by abdominal pain. Ultrasound indicated signs of torsion, and torsion of the right ovary was subsequently confirmed during laparotomy. A salpingo-oophorectomy was performed, and histological examination revealed a moderately differentiated Sertoli-Leydig cell tumor.

Sertoli-Leydig cell tumors often present with hormone-related or non-hormonal symptoms. Surgery plays a crucial role in both diagnosis and treatment. Postoperative treatment is not necessary for well-differentiated Sertoli-Leydig cell tumors in stage IA-IB. However, patients with grade 2–3 disease, advanced stage, or heterologous elements may consider adjuvant treatment.

As these tumors are rare, this case contributes to the documentation of Sertoli-Leydig cell tumors, with a case diagnosed due to ovarian torsion. The case highlights the importance of establishing international registries of Sertoli-Leydig cell tumor cases for standardized management.

1. Introduction

Torsion occurs as a complication in 10% of cases of ovarian tumors. It predominantly occurs in benign ones, particularly simple cysts, dermoid cysts and serous cystadenomas, while malignant tumors are less prone to torsion [1]. Sertoli-Leydig cell tumors (SLCTs) constitute a rare subtype of malignant ovarian tumors, accounting for less than 0.2% of all ovarian cancers. They are classified as a subgroup of sex cord stromal tumor (SCST), which represent approximately 7% of all malignant ovarian neoplasms [2]. This is a report of a case of ovarian torsion secondary to SLCT in a 39-year-old patient with an emergency presentation to hospital.

2. Case Presentation

A 39-year-old woman, gravida 1 para 1, presented to the emergency department with complaints of lower abdominal pain. It had begun 6 h earlier, localized to the right iliac fossa, and was not relieved by analgesic treatment. She had no associated symptoms such as nausea, diarrhea or fever. The patient's medical history included a thyroidectomy performed 16 years earlier, with ongoing hormonal treatment. She reported regular menstrual cycles lasting 5–6 days, with normal menstrual flow and no significant dysmenorrhea.

On admission, the patient was hemodynamically stable and afebrile. She presented tenderness and abdominal guarding upon palpation in the right iliac fossa. Ultrasonography revealed an enlarged right ovary, in a retro-uterine and medial location, measuring 75x82x53 mm. There was peripheral distribution of the follicles, with a stromal edema and peripheral vascularization on color Doppler, a typical whirpool sign of torsion, and no pelvic peritoneal effusion. The uterus and left ovary were normal. There was neither leukocytosis nor raised CRP. BHCG was negative.

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Due to the persistence of pain and the large size of the right ovary, the diagnosis of an ovarian torsion was made and an emergency laparotomy was performed. The ovarian torsion was confirmed during surgery. The right fallopian tube and ovary were twisted by 720 degrees, with a purple-black appearance on the surface of the right ovary (Fig. 1). Despite a careful detorsion, there was no significant change in the color of the ovary nor a decrease in edema. Right salpingo-oophorectomy was conducted. The postoperative course was uneventful, with resolution of pain after surgery. The patient was discharged in stable condition on the third postoperative day.

Histology revealed a moderately differentiated Sertoli-Leydig cell tumor of 9x7x6 cm with heterogeneous elements and absence of capsular rupture (Fig. 2). In immunohistochemistry, moderate positive staining was observed in Sertoli cells and intense staining in Leydig cells for antibodies against calretinin, inhibin, and CD56 (Fig. 3).

Tumor marker testing was then requested, including alpha-fetoprotein (AFP), cancer antigen 125 (CA125), beta-human chorionic gonadotropin (β-HCG), and inhibin B, which were found to be normal. As part of the staging evaluation, a thoraco-abdominopelvic CT scan was performed and was normal.

At a multidisciplinary meeting, the consensus was to offer adjuvant chemotherapy based on the histology results. However, after the decision was announced, the patient was lost to follow-up. She consulted 8 months later, and after another multidisciplinary meeting, it was decided to schedule radiological follow-up every 6 months.

3. Discussion

SLCTs, a highly unusual type of neoplasm, are classified as sex cord stromal tumors of the ovary [3]. They can manifest at any age, with reported cases ranging from 4 months to 81 years, but are mostly observed in second and third decades [2].Typically, SLCTs are localized to a single ovary, often the right side, bilateral occurrences being extremely rare, reported in approximately 1.5% to 2.0% of all SLCT cases [3].

There are three distinct categories of SLCTs, based on the compositions of Sertoli and Leydig cells: pure stromal tumors, pure sex cord tumors, and mixed sex cord-stromal tumors [4]. Many of these tumors demonstrate hormonal activity corresponding to their cell origin, and have the capacity to produce testosterone [5], often resulting in hyperandrogenism in adolescents [4], with hormonal-related symptoms including hirsutism, clitoromegaly, voice deepening, and acne [6]. 50% of SLCTs do not exhibit endocrine symptoms, and patients commonly present with symptoms such as abdominal discomfort or pain, ascites, or tumor rupture [3]. Pain is the most commonly reported symptom associated with SLCT [6]. In this case, the patient presented with pain but had no androgenic features. In the present case, postoperative tumor markers, evaluated after histopathological diagnosis, were within normal ranges, likely attributed to their evaluation timing post-surgery. Two recent studies have explored similar cases. One study by Lauren Roth et al. discusses an ovarian leydig cell tumor associated with recurrent torsion and virilization in an adolescent patient [4], while another by P.Pai et al. describes a case of sertoliform endometrioid tumor of the ovary presenting as torsion in a 55-year-old premenopausal woman [7].

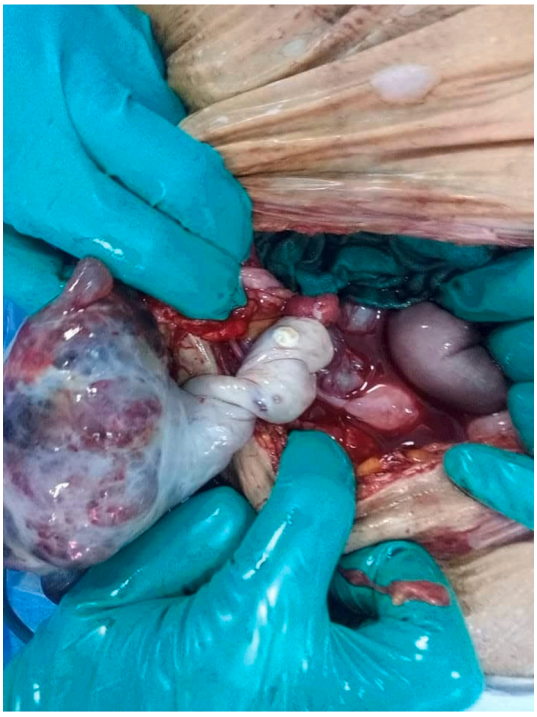

Fig. 1. Image of the right fallopian tube and ovary twisted by 720 degrees.

The radiological findings of SLCT are not extensively documented, as few studies have thoroughly described the imaging features of the disease. On ultrasonography, the most frequent presentation is a pure solid mass. However, a multicystic-multilocular appearance without any solid area is not uncommon [2]. These masses typically exhibit an average diameter ranging from 3 to 11 cm [3]. Additionally, the characteristic appearance of torsion on ultrasound is demonstrated by unilateral ovarian enlargement with edema and peripheral arrangement of the follicles. However, the use of color Doppler for diagnosing torsion remains controversial due to the dual blood supply of the ovary [8]. In the present case, ultrasonography revealed an enlarged right ovary with a multifollicular appearance, along with peripheral distribution of these follicles and peripheral vascularization on color Doppler, supporting the diagnosis of torsion.

Histologically, SLCTs are defined by the proliferation of Sertoli and Leydig cells in different proportions and are categorized as: well-differentiated, intermediate differentiation, and undifferentiated [6]. Heterologous elements are present in around 20% of SLCTs and can be divided into two types: endodermal elements, with the most common being gastrointestinal epithelium, and mesenchymal elements, such as cartilage or skeletal muscle [3].

Due to the rarity of SLCTs of the ovary, there have not been any randomized trials evaluating the effectiveness of adjuvant therapy [9]. Surgery is crucial for both diagnosis and treatment, with the approach tailored to factors like age, fertility needs, clinical stage, tumor size, and differentiation [2]. Preventing tumor spillage during surgery is critical as it is linked to higher recurrence risk [10]. Postoperative treatments may include chemotherapy, radiotherapy, or their combination based on tumor stage and differentiation [11]. For well-differentiated SLCTs in stages IA-IB, adjuvant therapy is not necessary, with fertility-sparing surgery sufficient after complete surgical staging due to favorable prognosis [9]. Patients with grade 2–3 disease, advanced stage, or heterologous elements may consider adjuvant treatment with bleomycin, etoposide, and cisplatin [9]. Limited studies suggest neoadjuvant chemotherapy for those with poor prognostic factors or intermediate/poor differentiation. Bleomycin, etoposide, and cisplatin are deemed effective as first-line chemotherapy [12]. In the present case, ovarian torsion constituted a therapeutic emergency, and considerations regarding the oncological approach were discussed in a multidisciplinary meeting after the surgical intervention.

DICER1 gene testing is recommended, given that more than 90% of SLCT patients exhibit DICER1 mutations [5]. These genetic alterations can lead to changes in microRNA expression, affecting ovarian tissue by disrupting the regulation of genes responsible for gonadal differentiation and cell proliferation [13]. Specifically, there is a downregulation of crucial genes for ovarian development, an upregulation of genes promoting Sertoli cell differentiation, and a suppression of CYP19A1, which diminishes aromatase activity and thus exerts an androgenic effect [5].

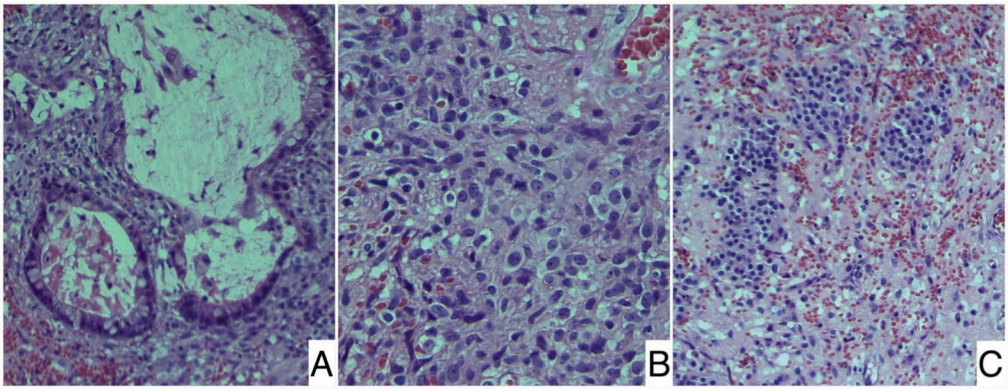

Fig. 2. : Histology images.
A: Gx20 Component of Sertoli and Leydig cell proliferation.
B: Gx40 sertoli cells.
C: Gx20 Leydig cells.

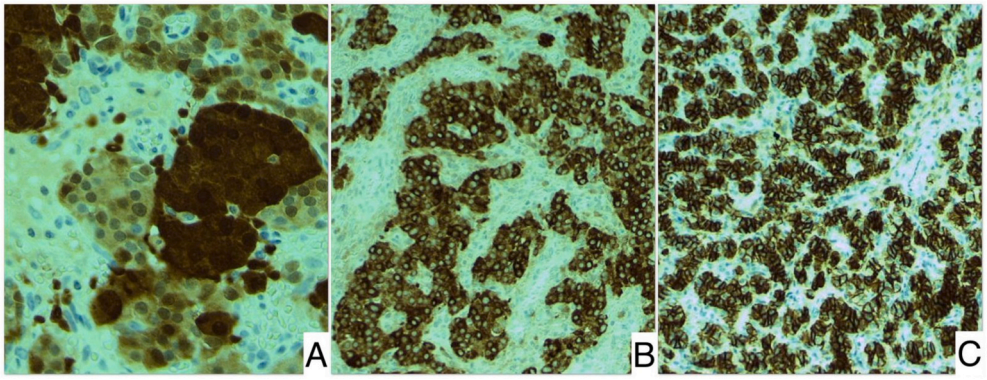

Fig. 3. : Immunohistochemistry images.
A: Gx40 positive for calretinin.
B: Gx20 strongly positive for inhibin.
C: Gx40 CD56 strongly positive in Leydig cells.

A diagnosis of SLCT should prompt referral to a clinical genetics service due to potential germline mutation and familial malignancy risk, with knowledge of a DICER1 mutation informing both current tumor management and future cancer risk assessment [13].

4. Conclusion

In conclusion, SLCT represents a rare histological subtype of ovarian sex cord-stromal tumor, and can present with hormone-related or non-hormonal symptoms. The primary treatment is surgical resection. Postoperative treatments may include chemotherapy, radiotherapy, or a combination thereof based on tumor stage and differentiation. Given the potential for recurrence, long-term follow-up is strongly recommended. Efforts should focus on establishing international registries of SLCT cases to enhance statistical power in future studies.

Contributors

Samia Tligui contributed to patient care, conception of the case report, acquiring and interpreting the data, undertaking the literature review and drafting the manuscript.

Hounaida Mahfoud contributed to the conception of the case report, acquiring and interpreting the data and revising the article critically for important intellectual content.

Samia Sassi contributed to drafting the manuscript, acquiring and interpreting the data and undertaking the literature review.

Hanane Inrhaoun contributed to drafting the manuscript, acquiring and interpreting the data and undertaking the literature review.

Najat Lamalmi contributed to undertaking the literature review and revising the article critically for important intellectual content.

Fatima El Hassouni contributed to undertaking the literature review and revising the article critically for important intellectual content.

Samir Bargach contributed to undertaking the literature review and revising the article critically for important intellectual content.

All authors approved the final submitted manuscript.

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Patient consent

Consent was obtained from the patient to publish the clinical details and the images included.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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