



## Case report

## Primary perimenarcheal ovarian leiomyosarcoma: A case report and review of the literature

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## ABSTRACT

**Introduction and importance:** Leiomyosarcomas of the ovary are extremely rare neoplasia usually occurring in perimenopausal patients.

**Case presentation:** A 16-year-old female patient, with no particular pathological history, who presented with chronic pelvic pain. On imaging; presence of a suprauterine solid cystic formation of 12 cm long axis. Tumor markers were normal.

On exploration, solid cystic formation of 15 cm long axis. A total hysterectomy with bilateral salpingo-oophorectomy associated with bilateral pelvic and para-aortic lymphadenectomy and a total omentectomy were performed. Anatomical pathology; an ovarian leiomyosarcoma.

**Clinical discussion:** Leiomyosarcomas of the ovary are extremely rare, representing less than 1% of all ovarian malignancies. They probably derive from the smooth muscle component. No definitive diagnostic criteria for ovarian leiomyosarcomas have been adopted to date, however the histological criteria adopted are those for uterine leiomyosarcomas. The differential diagnosis of these tumors includes fibrosarcomas, rhabdomyosarcomas, thecomas and extradigestive stromal tumors. The prognosis of ovarian leiomyosarcomas is generally poor.

Complete surgical resection remains for all authors the cornerstone of treatment.

The benefit of adjuvant therapies, namely chemotherapy or radiotherapy, remains to be proven.

**Conclusion:** Leiomyosarcoma of the ovary, although exceptional, should always be included among the diagnostic possibilities when an ovarian cyst of organic appearance is discovered in a perimenarcheal patient.

### 1. Introduction

Leiomyosarcomas of the ovary are extremely rare neoplasia usually occurring in perimenopausal patients [1]. They represent less than 1% of malignant tumors of the ovary [2] with less than 30 cases documented in the literature. We report here the third case of ovarian leiomyosarcoma occurring in perimenopause, after the one published by Saïm et al. in 2006. This work has been reported with respect to the SCARE 2020 criteria [3].

### 2. Case report

The patient was 16 years old. She had her first menstrual period at the age of 12 and a half and her cycles were irregular, without any particular pathological history. The patient complained of chronic

pelvic pain, going back six months without digestive or urinary signs. Clinical examination revealed abdomino-pelvic distension with the presence of a mass reaching the umbilicus. Ultrasound examination revealed a suprauterine mass with irregular solid-cystic contours, multi-compartmentalised with a predominance of the solid part, measuring 12.59 cm in long axis, varies not seen, and a moderate amount of peritoneal effusion (Fig. 1).

Abdominal-pelvic CT: Pelvic solid-cystic formation of 15 × 9.3 cm with intraperitoneal effusion of medium abundance visible in the perihepato-splenic area and in the parieto-colonial gutter. There was no deep adenopathy (Fig. 2).

Tumor markers CEA, CA125, AFP and β-HCG were normal.

Laparoscopy was performed. Pelvic exploration found a 15-cm large solid-cystic looking right ovarian cyst with a large solid component.

A laparoconversion was performed and intraoperatively, a right

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**Fig. 1.** A suprauterine mass with irregular solid-cystic contours, multiply partitioned with predominance of the solid part measuring 12.59 cm in long axis. Ovaries not seen. Medium-sized peritoneal effusion.



**Fig. 2.** Pelvic solid-cystic formation of 15 × 9.3 cm with a moderate amount of intraperitoneal effusion visible in the peri-hepato-splenic area and in the parieto-colonic gutter. Absence of deep ADP.

ovarian mass with an irregular surface, adherent to the douglas. A total hysterectomy with bilateral salpingo-oophorectomy associated with bilateral pelvic and para-aortic lymphadenectomy and total omentectomy were performed.

Frozen pathology results showed that the tumor was a malignant mesenchymal tumor. The omentum, uterine serosa, and pelvic lymph nodes were infiltrated by the tumor. Immunohistochemistry at magnification 20 (a) and magnification 40 (b) yields diffuse expression of AML by the tumor cells in favor of ovarian leiomyosarcoma (Fig. 3).

According to the 2014 FIGO classification, the final histopathologic diagnosis was stage IIIC ovarian leiomyosarcoma. The patient was treated with 6 courses of adjuvant chemotherapy including 50 mg/m<sup>2</sup> doxorubicin and remained unharmed at the 23-month follow-up.

### 3. Discussion

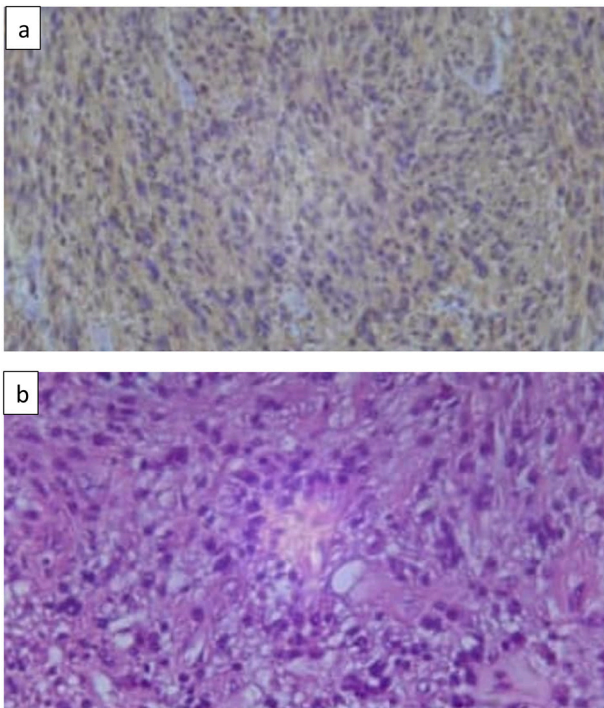
Leiomyosarcomas of the ovary are extremely rare, representing less than 1% of all ovarian malignancies [2]. The rarity and heterogeneity of

this pathology have made its classification difficult. The diagnosis, treatment and prognosis also remain uncertain [4]. Although these lesions have been reported in patients between 17 and 84 years of age, it has often been accepted that this is a postmenopausal pathology [1,5]. Only three of the 30 cases published in the literature occurred in patients younger than 35 years of age [4], two of which were perimenarcheal.

The case we report is the third, after the one published by Saïm et al. in 2006 [6], occurring in a perimenarcheal patient.

Very few etiological or predisposing factors are known to date [7]. Leiomyosarcomas of the ovary probably derive from the smooth muscle component, which is present in the blood vessel wall, in the cortical stroma around the follicles and corpus luteum or in the ovarian ligaments at their insertion to the ovary [4]. The smooth muscle component of ovarian teratomas has also been suspected as a potential source of these tumors [8,9].

To date, no definitive diagnostic criteria for ovarian leiomyosarcomas have been adopted [5]. The histological criteria adopted are those for uterine leiomyosarcomas. According to Lerwill et al. [10], in cases



**Fig. 3.** Immunohistochemistry at magnification 20 (a) and magnification 40 (b) shows diffuse expression of AML by tumor cells in favor of ovarian leiomyosarcoma.

where the distinction between benign and malignant smooth muscle tumor of the ovary remains difficult, the finding of more than five mitoses per ten fields at high magnification and the presence of significant cytonuclear atypia are sufficient arguments to make the diagnosis of leiomyosarcoma of the ovary, even in the absence of foci of tumor necrosis [5].

The differential diagnosis of these tumors includes fibrosarcomas, rhabdomyosarcomas, thecomas and extradigestive stromal tumors. The immunohistochemical study is therefore of great importance, showing the positivity of leiomyosarcomas to actin and vimentin [4].

The prognosis of ovarian leiomyosarcomas is generally poor. Several immunohistochemical markers have been studied in the hope of collecting prognostic factors for these malignant tumors.

Indeed, Mayerhofer et al. [1] found that 90% of tumor cells express P53 and 30% express Ki67. The latter is a protein known to be an indicator of cell proliferation and therefore of tumor aggressiveness.

Thanks to their studies, Mayerhofer et al. [1] have also defined other immunohistochemical parameters of leiomyosarcomas. Indeed, they have shown that these tumors are positive for the matrix metalloproteinases MMP1 and MMP2. The latter, as well as an angiogenic factor called VEGF detected in these ovarian sarcomas, are factors that predict the aggressiveness of the tumor and therefore its subsequent prognosis [1].

In a second study, Mayerhofer et al. also demonstrated high tumor positivity to BCL2 with more than 80% of cells positive. This protein is involved in cell cycle regulation. It is also able to promote cell replication by reducing the need for growth factors and thus plays an important role in tumor growth [1].

Recommendations for the optimal management of these tumors are still very difficult to make. However, complete surgical resection remains for all authors the cornerstone of treatment. Classically, the surgical approach is medial. However, there are always exceptions or marginal indications.

In our case, we are dealing with a 16 year old girl with a 15 cm tumor formation, normal tumor markers. Laparoscopy was not worrisome.

Median laparotomy seems to us to be the correct attitude in this case, allowing to perform all the necessary procedures in front of a tumor suspected of malignancy.

The benefit of adjuvant therapy, i.e. chemotherapy or radiotherapy, has yet to be proven [2,4]. In our case, because the omentum, uterine serosa and pelvic lymph nodes were infiltrated by the tumor, the patient received 6 courses of adjuvant chemotherapy including 50 mg/m<sup>2</sup> doxorubicin and remained unharmed at the 23-month follow-up.

#### 4. Conclusion

Leiomyosarcoma of the ovary, although exceptional, must always be included among the diagnostic possibilities when an ovarian cyst of organic appearance is discovered in a perimenarcheal patient.

The prognosis of this pathology is generally unfavourable. Recently, markers such as Ki67, metalloproteinases MMPs and VEGF are parameters that can predict the aggressiveness of these tumors. Further case analysis and long-term follow-up of the identified cases is necessary to confirm the prognostic value of these parameters and thus contribute to a better approach to the prognosis of patients with this pathology.

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#### Ethical approval

I declare on my honor that the ethical approval has been exempted by my establishment.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

Vilaly Khadjetou: writing the paper.

Telmoudi Ely Cheikh: Corresponding author writing the paper and operating surgeon.

Med Vadel Ahmed Haiba: study concept.

Med Ahmed Ahmed Cheikh: study concept.

Boukhary Nadi Mouhamed: study concept.

Bonahy Abdi Ahmed: correction of the paper and operating surgeon.

#### Registration of research studies

None.

#### Guarantor

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#### Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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