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Case Report

Sertoli-Leydig tumor in a 17-year-old girl: a case report

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ABSTRACT

Sertoli-Leydig tumors of the ovary are rare. They can affect women in all age groups but are more frequent in the first three decades. No case of Sertoli-Leydig cells tumors has been reported in the Haitian literature. We present a case of a 17-year-old teenage who was seen in the outpatient clinic of the Mirebalais teaching hospital for: amenorrhea, change in voice and hair loss. After our evaluation and the completion of certain assessments, the clinical impression of a stromal and sexual cord tumor was retained, which guided us to perform a laparotomy. We conclude that in the management of this type of case, it is important to take into account the desire to preserve the patient's subsequent fertility and if there is one point that should not be overlooked, it is the psychosocial repercussions of the pathology before and after its treatment.

Keywords: Case report, Tumor, Sertoli-Leydig, Ovary, Haiti

INTRODUCTION

Sertoli-Leydig tumors of the ovary are tumors made up of variable proportions of Sertoli cells and undifferentiated stromal cells.[1] In fact, they represent approximately less than 0.5% of all ovarian tumors, they can affect women in all age groups from 2 to 75 years old however are more frequent in the first 3 decades.[2,3] The clinical presentation of these tumors varies considerably and can range from an asymptomatic form to a clinical profile of extreme virilization.[2] About half of patients with Sertoli-Leydig tumors present with endocrine manifestations such as virilization.[4] These signs of virilizations can be amenorrhea, hirsutism, male pattern of pilification and these characteristics are due to an increase production of androgens by the tumor cells.[2]

Radiological images of such tumor are not well defined and remain to be elucidated, and histopathological examination will confirm the diagnosis.[2] Most of these tumors are unilateral and their preferred treatment is surgery while taking into account the desire to preserve fertility.[2] The management of these tumors remains difficult due to their rarity and the lack of a standard protocol.

In Haiti, data are not reported in the medical literature concerning cases of Sertoli-Leydig tumors, even less in the age group relating to adolescents. In this context, we present here the case of a seventeen-year-old girl who was treated for a Sertoli-Leydig cell ovarian tumor with the signs of virilization. The objective of this article is to draw attention to this rare case encountered in a teenager at the Mirebalais teaching hospital, to highlight the importance of the diagnostic procedure for the tumor and its management based on the Haitian context, and its associated psychosocial repercussions.

CASE REPORT

A 17-year-old teenager, virgin, and schoolgirl, came to the outpatient gynecology clinic at the Mirebalais teaching hospital, Haiti for: ten months of amenorrhea, change in voice and hair loss. Because of the signs of virilization, the

patient was the victim of mockery from her classmates. People around her teased her about her voice to the point that she spoke less. This pushed patient to a social withdraw.

She had no significant past medical, surgical, or family history. She did not use alcohol, tobacco or illicit drugs. On physical examination, her vital signs were within normal limits, the hair was short, she had hirsutism. The abdomen was soft and depressible with no evidence of palpable masses. Examination of the vulva reveals clitoromegaly (Figure 1A). On abdominal ultrasound the right ovary was replaced by a mixed mass, with regular contour, well delimited, and measured approximately 6.23 cm by 4.05 cm (Figure 1B). Uterus, fallopian tubes, left ovary and kidneys were unremarkable. In Table 1, we described the hormonal profile and the tumor markers that we performed for the patient.

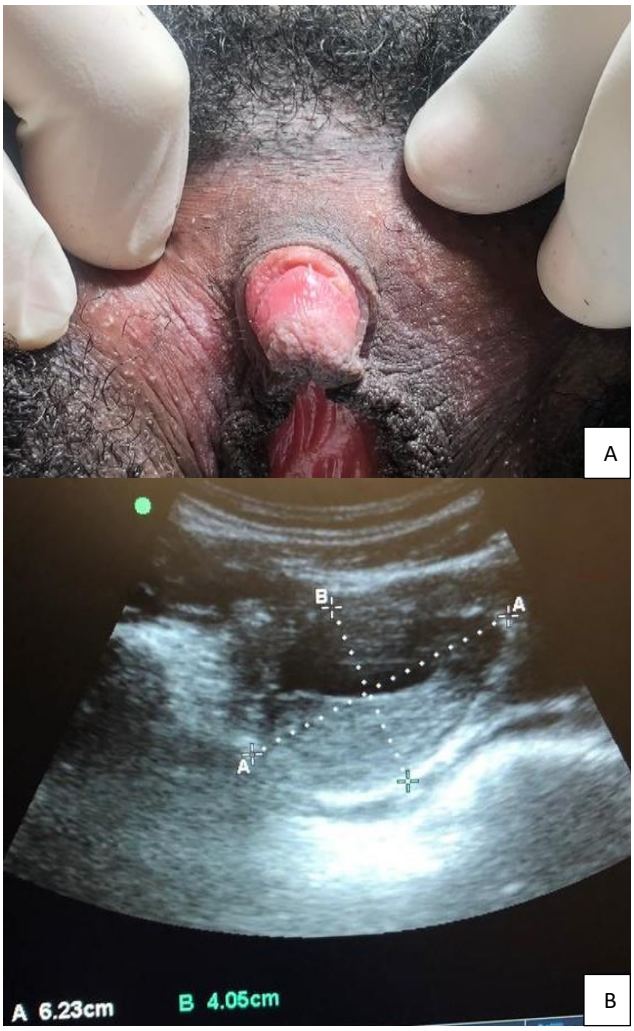

Figure 1 (A and B): Demonstration of clitoromegaly and visualization of the mass on ultrasound.

After discussing the likelihood of sex cord stromal tumor diagnosis and its management with the patient and her parents, their biggest concern when making a decision was about her fertility, then came worry about her voice, if it will come back as before after surgery.

Before the laparotomy, the patient received psychosocial support to prepare her for surgery and for the management of her psychological symptoms. At the laparotomy, we found a right ovarian mass with two consistencies (soft and a solid part), with a smooth surface, without adhesions with the neighboring structures (Figure 2A). The uterus, fallopian tubes, left ovary (Figure 2A), and the omentum were macroscopically unremarkable. We performed a right oophorectomy (Figure 2B). After advice on her post-operative follow-up and psychological consultation, the patient was discharged after three post-operative days without complications.

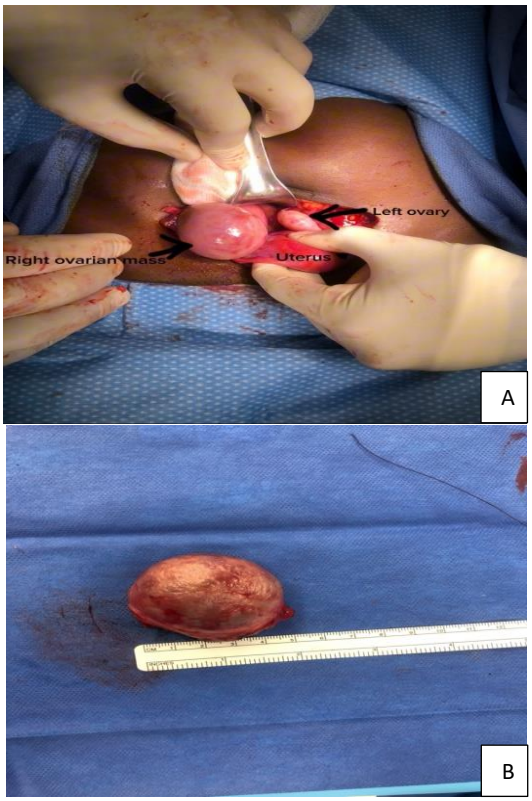

Figure 2 (A and B): Mass in per op and right ovarian mass after resection.

During the follow-up, one month after the intervention, the patient had a return of menstruation, a slight improvement in the voice, a reduction in hirsutism and the testosterone level dropped to 1.03 ng/ml (normal value: 0.1-0.6 ng/ml). Given this clinical and biological improvement, this reinforced our initial clinical impression while waiting to have the histopathological results which were not yet available at this time.

Three months after surgery, we received the results of the specimen. On microscopy, a solid zone rich in Sertoli cells organizes into a cord and tubules between which Leydig cells insinuate and a cystic zone containing glandular formations coated with a mucosecretant intestinal

epithelium, on the periphery of this zone we observe Sertoli cells and the stroma is edematous (Figure 3). The pathologist diagnosed a moderately differentiated Sertoli-Leydig tumor.

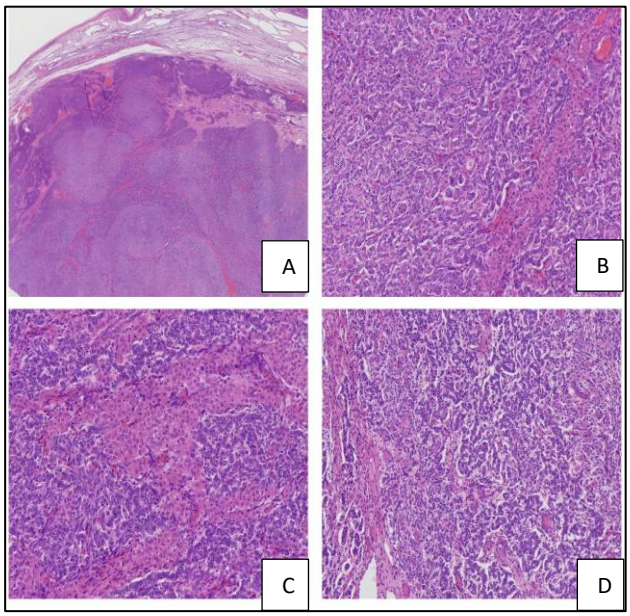

Figure 3 (A-D): Pathological slides of the tumor.

Table 1: Hormonal finding and tumor markers.

Tests	Unit	Results	Normal values
FSH	mIU/ml	1.3	1-6.5
LH	mIU/ml	7.17	7-12.9
Prolactin	ng/ml	17.44	1.9-25.9
Estradiol	pg/ml	55	13-300
Testostérone	ng/ml	5.99	0.1-0.6
CA 125	U/ml	17.37	0-35
CA 19-9	U/ml	11.37	0-36
CEA	µg/l	1.8	0-5
LDH	U/l	180	150-450

DISCUSSION

Sertoli-Leydig tumors can affect patients in any age group, 51% of cases occur in the first 3 decades of life.[5] Other data show that it is more common in the first 2 decades of life, as in this 17-year-old girl. The signs and symptoms of Sertoli-Leydig tumors are due to testosterone production on one side and abdominal mass on the other.[2,6] Patients typically experience hormonal changes, such as signs of virilization that usually precede anovulation or amenorrhea, which was exactly the case for our patient.[2,7]

These tumors don't only have physical repercussions; we can also observe changes in psychosexual behavior in 30 to 50% of cases.[1] Although typical cases of the psychosocial effect of Sertoli-Leydig tumors at this age have not really been described, some cases could be found in a different age group including a 36-year-old woman where the signs of virilization had psychosocial effects with a state of stress linked to the desire for fertility, and loss of fertility in relation to surgery, and also social pressure, judgment from those around him.[8] Indeed, the primary concern of the teenager was also to know if she will be able to give birth in the future. In addition to fertility considerations, hirsutism and hoarseness can have serious psychosocial consequences for affected women, and many epidemiological studies have shown that affected women suffer significantly more often from mental illnesses such as depression and anxiety disorders.[9] In the case of the patient, there was a social withdrawal, with a withdrawal into herself due to the various mockeries of her classmates showing the psychological effect in our case.

Transvaginal ultrasound remains the imaging modality of choice for the first evaluation of adnexal masses because of the high sensitivity, its relevance and the cost, however, being a virgin, the abdominal route was used.[10] These tumors generally appear on ultrasound as a heterogeneous vascularized tissue mass with solid areas associated with anechoic liquid areas.[1] In this case, we had a unilateral mass of heterogeneous echostructure with mixed component. Other imaging studies can be used for a better characterization of the ovarian mass such as CT, MRI but unfortunately, we do not have these studies available for our patient.[11] For these patients with Sertoli-Leydig tumour, the assessment also depends on the symptomatology. In this context, patients showing signs of virilization, the dosage of the following main major androgens: testosterone, delta-4-androstenedione, dehydroepiandrosterone, must be done.[1] For patients with amenorrhea, it's coupled with the dosage of pituitary hormones such as FSH, LH, and Prolactin.[1] In our context, only testosterone could be measured, which was nearly ten times higher than normal and the other hormone dosage was within normal limits. However, in the diagnostic approach, other markers were performed such as CA 125, CA 19-9, CEA and LDH, which were unremarkable.

The management of Sertoli-Leydig tumors remains difficult, mainly because of the lack of a standard protocol.[11] However, the surgical approach is most often prioritized. Generally, these Sertoli-Leydig tumors are unilateral and limited to the ovary, and are mainly diagnosed at an early stage.[3] Then, an adnexectomy or an oophorectomy is most often performed.[1] In this context, our patient thus benefited from conservative management with right oophorectomy. For cases with no desire for fertility, the treatment is a total hysterectomy with bilateral adnexectomy and can be combined with an omentectomy.[1] As another approach, lymph node dissection and adjuvant chemotherapy as needed is modeled on malignant tumors that have a poor prognosis.[1] A biopsy of the omentum and peritoneal washing could have been performed, but given the age of the patient and the absence of macroscopic features of these structures during the surgery, the team omitted these steps.

On microscopy these tumors present with areas of uncontrolled proliferation of varying degrees of differentiation of tubules lined with Sertoli cells and intermediate nests of Leydig cells.[1,11] Well and moderately differentiated histological variants are the most common.[11] It should be noted that heterologous elements can be found in these last two forms.[1] It has also been reported that the most frequently found heterologous element is the benign gastrointestinal type epithelium.[3,12] For the case presented here, we found a solid zone rich in Sertoli cells organized in cords and tubules between which Leydig cells insinuate, and the heterogeneous element found was a mucosecretant intestinal type epithelium. The only concern is the tumor was moderately differentiated which can justify the adding of bleomycine, ectoposide and cisplatin (BEP) or paclitaxel plus carboplantin (PC) as adjuvant chemotherapy in the treatment plan. As the pathological results were ready too late the team continues with the expectative management consisting in clinical, biological and radiological evaluation. We plan to follow this patient longer than expected as she did not receive adjuvant chemotherapy. This case delineates the necessity to train a comprehensive number of pathologists in the developing world to avoid such delay, as only one full time pathologist was available for the whole hospital.

CONCLUSION

Sertoli-Leydig tumors are very rare. The diagnosis, although difficult, was made with clinical data such as amenorrhea, and signs of virilization, associated with testosterone levels and ultrasound. For the management, although several modalities have been described, but considering the parameters of our patient, we performed an oophorectomy. A multidisciplinary approach is important for the management of this type of case. The development of radiological and histopathological diagnostic capabilities could improve the care of these patients in developing countries.

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