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Case Report

Hyperandrogenism of Tumor Origin in Adolescence: A Case Report

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Abstract

A case is presented of hyperandrogenism with signs of virilization in an adolescent female diagnosed with ovarian Sertoli-Leydig cell tumor and subjected to laparoscopic unilateral adnexectomy, with immediate normalization of the laboratory test findings and progressive clinical improvement following surgery.

Keywords: Hyperandrogenism; Virilization; Adolescent; Sertoli-Leydig cell tumor; Adnexectomy; Laparoscopy

Introduction

A 12-year-old girl presented with deepening and cracking of the voice, marked acne on the face and upper part of the dorsal region, increased body hair predominantly in the genital zone and buttocks, and the absence of menstruation over the last two months.

Case Presentation

Her previous pubertal development had been normal, with menarche in October 2021, and no sexual intercourse. In view of the clinical picture, Pediatrics requested blood testing that revealed increased concentrations of 17-OH-progesterone (7.03 ng/ml), testosterone (3.35 ng/ml) and androstenedione (6.04 ng/ml). An ACTH test to assess a possible adrenal gland origin of the hyperandrogenism proved negative. Abdominal ultrasound in turn revealed a normal left ovary and uterus; the right ovary could not be visualized.

In view of the above findings, the patient was referred to Gynecology and a Magnetic Resonance Imaging (MRI) study was carried out. The gynecological exploration showed a clear increase in body hair in the region of the midline line alba, external genitals and perineal region, together with hypertrophy of the clitoris.

Trans rectal ultrasound showed a uterus of normal size and with a thin endometrium. The left ovary was increased in size at the expense of a solid-cystic tumor mass with a low-intensity Doppler signal (Figure 1). The right ovary appeared normal (Figure 2).

Based on the abovementioned ultrasound, clinical and laboratory test findings, a left ovarian Sertoli-Leydig cell tumor was suspected as the cause of hyperandrogenism and secondary virilization of the patient. The MRI findings were consistent with the suspected diagnosis and showed an enlarged left ovary due to the presence of a tumor mass (4.8 cm × 3.6 cm × 3.7 cm). The lesion was well delimited, with a smooth surface and a heterogeneous internal signal intensity showing irregular cystic areas measuring up to 2.2 cm in size and a low signal intensity solid component suggestive of a fibrous stromal mass.

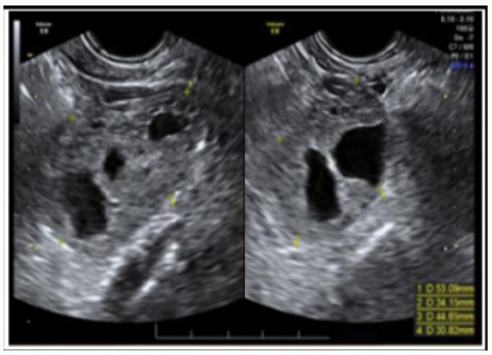

Figure 1: Ultrasound view of the left ovary, enlarged in relation to the right ovary and containing solid and cystic areas.

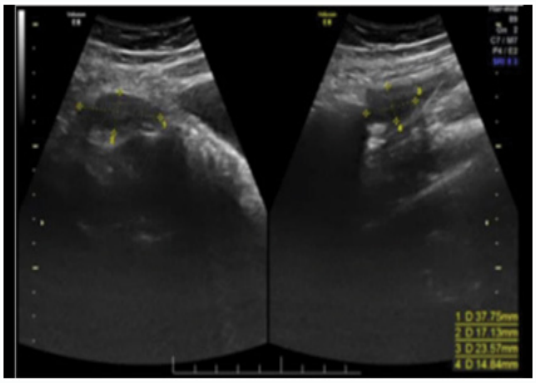

Figure 2: Ultrasound view of the right ovary, of normal dimensions.

The tumor marker tests proved normal, and a thoracic-abdominal-pelvic computed tomography scan discarded loco regional as well as distant tumor spread. After evaluation of the case by Pediatric

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Oncology, surgical treatment was decided. The advisability of left oophorectomy versus laparoscopic left adnexectomy was commented with Pathology, and following a review of the literature on the subject and in order to allow a better analysis of tumor spread, unilateral adnexectomy was decided.

Surgery revealed a normal uterus. Both ovaries were oval in shape, with a smooth pearly surface, and the left ovary was larger than the left ovary (approximate maximum diameter 5 cm and 3 cm, respectively). Both fallopian tubes appeared normal. The rest of the abdominal cavity showed no alterations (Figure 3).

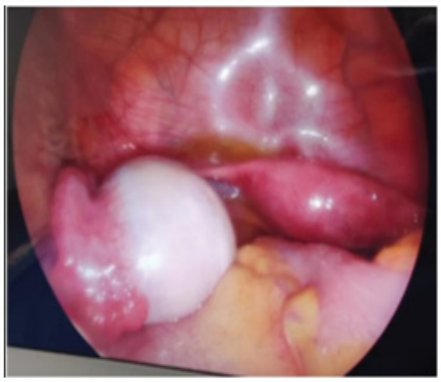

Figure 3: Laparoscopic view showing a uterus of normal appearance and size. Enlarged left ovary with a smooth and pearly surface.

The histopathological study of the surgical piece confirmed the diagnosis of moderate histological grade (G2) (WHO classification) ovarian Sertoli-Leydig cell tumor. The pathological classification (pTNM, AJCC 8th ed.) corresponded to pT1a (FIGO IA): tumor limited to the ovary and with an intact capsule. Disease-free surgical margins and fallopian tube.

With the results obtained, the case was discussed by the Pediatric Oncology Committee of the corresponding reference hospital, and the decision was made to not indicate adjuvant therapy. Controls were therefore scheduled in the outpatient clinic. The patient presented immediate normalization of the laboratory test findings and progressive improvement of the signs of virilization following surgery.

Discussion

Sertoli-Leydig cell tumor (androblastoma or arrhenoblastoma) is an uncommon disorder in clinical practice [1], hence the importance of reporting cases such as that described in this study. These lesions are included among the ovarian stromal tumors (5% of all ovarian tumors) and represent approximately 0.2% to 0.5% of all ovarian neoplasms [1,2]. They are characterized by the presence of cells derived from the sex cords (Sertoli cells) and cells derived from the stroma Leydig cells and fibroblasts), in variable proportions within the same tumor [3].

These tumors may manifest at any point in life, but are more common in women under 40 years of age (75% of all cases), as in our patient. The lesions are typically unilateral, with bilateral presentations accounting for only 5% of all cases [4].

Sertoli-Leydig cell tumors are characterized by increased androgen production, with the most common finding being plasma testosterone elevation (at least 2.5 times the upper limit of normal). Most lesions are hormonally active (70% to 85%). The clinical manifestations include signs and symptoms of virilization, such as a deepened voice, anomalous body hair distribution, hirsutism, a hypertrophic clitoris, and menstrual disorders in the form of chronic an ovulation (as in our patient) and infertility [4]. Surgical removal of the tumor usually results in normalization of the laboratory test parameters and the restoration of female characteristics [5].

Macroscopically, these are solid yellowish tumors measuring an average of 13 cm in long diameter [1]. In our case, however, the lesion measured 5 cm in size and was of a pearly white color with a smooth surface. The main prognostic factor is disease stage at the time of diagnosis [6]. Fortunately, the great majority of these tumors (92%) is confined to the ovary (stage I) and present a good prognosis. This allows for conservative management (unilateral adnexectomy), with the preservation of future fertility [4]. In elderly women hysterectomy and bilateral adnexectomy are indicated [3]. Between 3% to 20% of all cases are diagnosed in advanced stages, with pelvic and abdominal (but not distant) spread [4], requiring pelvic radiotherapy and chemotherapy [3].

In addition to disease stage at the time of diagnosis, other important prognostic factors are patient age and the histological differentiation grade of the tumor [6]. Adequate tumor typing by the pathologist is therefore crucial.

In our case, since the tumor was confined to the ovary (stage I) and exhibited moderate differentiation (G2), management was limited to surgery, with no need for adjuvant therapy. The 5-year survival rate is 70% to 90%. Recurrence is infrequent, typically occurs early (within less than 3-5 years), and has a poor prognosis. Close patient monitoring is therefore required during the first years after treatment.

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