Ovarian Sclerosing Stromal Tumour Associated with Hirsutism in an Adolescent

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Sclerosing stromal tumours are rare ovarian neoplasm in adolescent girls. Herein, we describe a case of 17 year-old adolescent girl presented with hirsutism and abdomino-pelvic palpable mass. Ferriman Gallwey score was 9 and abdominal examination revealed a pelvic mass in 150 mm in diameter. At laparatomy there was minimal free fluid in the pelvis and yellow to pink in colour pelvic mass in right ovary. Unilateral salpingo-oophorectomy was done. Histological examination reported as benign sclerosing stromal tumour of the right ovary. In adolescent patients sclerosing stromal tumours of ovary may be presented with hirsutism and abdomino-pelvic mass. Attentive pelvic examination and imaging is important in hirsute adolescent patients.

Keywords: Androgens, Hirsutism, Ovary

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Introduction

Sclerosing stromal tumours (SSTs) arise from ovarian stroma and are rarely associated with estrogen and, in exceptional cases, androgen secretion. SSTs are seen predominantly in the second and third decades of life. The common presenting clinical symptoms are pelvic pain, hypermenorrhea and menstrual irregularities. We present a case of SST of the ovary diagnosed in a hirsute adolescent girl. Hormonal activity and hirsutism with tumour are striking aspects of this case.

Case Report

A 17 year-old adolescent girl was complained of hirsutism for 6 months' that mainly affects abdominal region. Ferriman Gallwey score was 9. There was no clitoral enlargement and balding. Abdominal examination revealed a pelvic mass in 150 mm in diameter. Her body mass index was 23.4 kg/m^2. She had had menarche at 13 years old and has infrequent menstrual cycles (oligomenorrhea). On transabdominal ultrasonography, uterus was 40x30x20 mm in diameters; endometrial thickness was 12 mm with regular borders. In the right adnexial region, there was a 150x120x70 mm, solid mass with irregular borders. Left ovary was 30x20x20 mm and containing 5-6 antral follicles. No polycystic view was detected with ultrasonography. Minimal free fluid was found in pelvis. CA 125 level was 30.4 U/ml, CA 19-9: 20.8 U/ml and CA 15-3:12.4 U/ml which were all within normal limits. The laboratory findings showed mild elevation of testosterone and 17-hydroxy progesterone levels which were 98 ng/dL (normal range, 11 to 80 ng/dL) and 1.90 ng/mL (normal range, 0.1 to 1.08 ng/mL), respectively. Androstenedione and dehydroepiandrosterone-sulfate (DHEA-S) levels were 2.60 ng/mL (maximum level 3.08 ng/mL) and 3242 ng/mL (normal range, 1330 to 4410 ng/mL) (Biosource, Nivelles, Belgium). At laparatomy there was minimal free fluid in the pelvis, and uterus and left ovary were normal. One hundred and fifty millimetres mass, which was yellow to pink in colour and had a smooth and well-encapsulated surface, was seen on the right ovary (Figure 1). Unilateral salpingo-oophorectomy was done. Frozen section was reported as a benign SST of the right ovary. She was discharged from the hospital with good condition at postoperative day 2.

Microscopic examination of the right ovary showed cellular portions with fibroblasts, rounded vacuolated cells and prominent vessels as well as edematous and collagenous hypocellular areas (Figure 2). Immunohistochemical studies showed that the tumour was positive for smooth muscle vimentin, actin, inhibin and CD 34 but negative for S-100 protein, cytokeratin and desmin (Ultraview Universal DAB, Ventana Bench XT, USA)

Six months after the operation, despite the mild decrease in hormone levels (Testosterone: 67 ng/dL, Androstenedione:

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1.27 ng/mL, DHEAS: 3127 ng/mL.), hirsutism score was found to be unchanged.

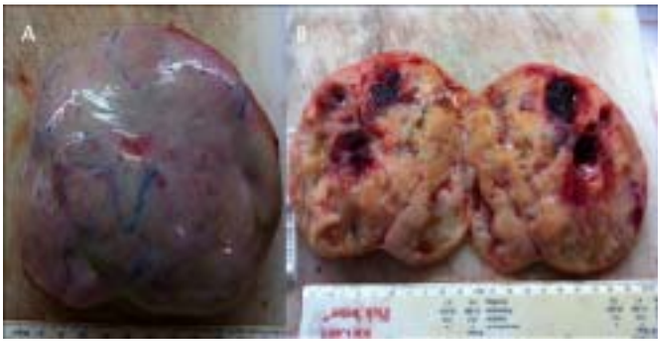

Figure 1: A: Macroscopy of right ovary, B: Macroscopic view; inside of the tumour

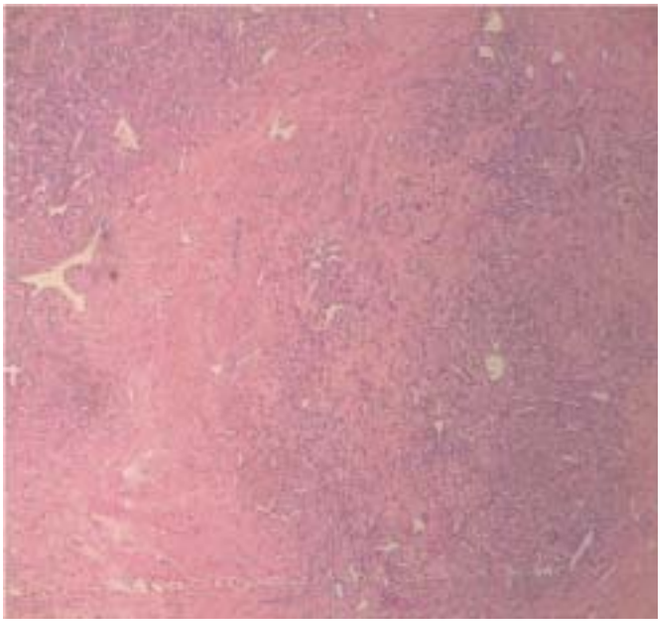

Figure 2: Microscopy of right ovary, marked sclerosis with edematous stroma (H & E, x 100).

Discussion

SSTs are an extremely rare and distinctive sex-cord stromal tumour. It was first described in 1973 by Chaldvardjian and Scully.[1] Gee and Russell showed clinical and biochemical markers of hormonal activity.[2] STTs have been only occasionally associated with estrogen and rarely androgen secretion. Some patients have presented with anovulation or masculinisation which resolved spontaneously once the tumour was removed.[3] Damajanov et al. showed decreasing estradiol and androgen concentration after tumour excision.[4] It can be associated with endometrial cancer but the causal relationship is not proven.[5] All SSTs reported to date have been benign and hormonal activity may be present but recurrence has not been described.[6]

In the present case, SST was associated with hyperandrogenic symptoms such as hirsutism without clitoral enlargement and balding. The levels of testosterone, andostenedione and DHEAS were mildly elevated. Though no cut- off values were established with regard to androgen levels causing hyperandrogenic symptoms, in the present case, the lack of other factor that potentially may lead to hyperandrogenism strongly suggest causal relationship between SST and clinical findings. Despite the persistence of hirsutism, her androgen levels were less than preoperational evaluation which implies complicated relationship between hormonal activity, target tissue response, and clinical outcomes that occurs in SSTs.

Despite the existence of a case with SST that shows elevated levels of CA125 in the literature, there is no specific marker that can be related with SSTs in clinical use.[7]

On sonograms the appearance of SSTs may be suspected to be malignant ovarian tumours because they show a mixed pattern with cystic and solid components.[8]

The differential diagnosis of SSTs of the ovary includes other sex-cord stromal tumours such as fibromas and thecomas. These may be differentiated from the SST on the basis of histopathology and immunohistochemical findings. Tiltman suggested that STTs and thecomas shares morphologic features and many antigenic determinants like smooth muscle actin, vimentin and thus are probably closely related entities.[9] Although overlap exists between the stromal tumours on the basis of immunohistochemistry and some morphologic features, distinctive clinical and pathological features of the STT almost always allows a specific differential diagnosis.

In conclusion, SSTs can affect adolescent girl with hyperandrogenism, and in the presences of adnexial mass, it should be considered in differential diagnosis of hirsutism.

Adolesan Bir Olguda Hirsutizm ile Birliktelik Gösteren Ovaryen Sklerozan Stromal Tümör

Sklerozan stromal tümörler adolesanlarda nadir olarak görülen neoplazmalardır. Bu yazıda hirsutizm ve abdomino-pelvik kitle ile başvuran 17 yaşında bir adolesan hasta sunulmaktadır. Ferriman Gallwey skoru 9 olan hastanın abdominal muayenesinde 150 mm büyüklüğünde pelvik kitle saptandı. Yapılan laparatomide pelviste minimal serbest sıvı ve sarı-pembe renkte sağ over kaynaklı kitle görüldü. Tek taraflı salpingooferektomi yapıldı. Histopatolojik inceleme sağ ovaryen benign sklerozan stromal tümör olarak raporlandı. Adolesan hastalarda overin sklerozan stromal tümörleri hirsutizm ve abdomino-pelvik kitle şeklinde prezante olabilir. Hirsutizm ile başvuran adolesan hastalarda dikkatli pelvik muayene ve görüntüleme önemlidir.

Anahtar Kelimeler: Androjenler, Hirsutizm, Over

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