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Case Report

Steroid Cell Tumour - A Rare Cause of Hirsuitism in a Female

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Abstract

Steroid cell tumours of the ovary account for < 0.1 % of all the ovarian tumours. It is a functioning sex cord stromal tumour. Previously designated as lipoid cell tumours, one-third of these tumours are considered malignant with the mean age of presentation at around 40 years. We present a case of 20 year old unmarried girl with regular cycles who came with sudden onset amenorrhea, hirsuitism, abdominal distension and signs of virilizationof 5 month duration and recent onset of dyspnea to Obstetrics and Gynaecology outpatient department. Moderate ascites was present. Clinical and radiological evaluation revealed a right adnexal mass with elevated serum testosterone. She was diagnosed with right ovarian benign functioning tumour and underwent right sapling oophorectomy. Histopathology confirmed the diagnosis. 2 weeks post operatively her testosterone levels decreased. Surgery is the treatment of steroid cell tumours although medical therapy using Gonadotrophin Releasing Hormone [GnRH] analogues has been tried recently in recurrent or inoperable cases.

Introduction

Steroid cell tumours arerare functioning stromal ovarian tumour accounting for <0.1% of all ovarian tumours.[1] One of the subtypes, not otherwise specified accounts for one half of the steroid cell tumours. The most common occurrence is around 43 years of age[2,3]. The symptoms are amenorrhoea, virilising symptoms like hirsuitism, change in voice pattern etc. Ascites is a rare finding. About one third of the steroid cell tumours are malignant. Surgery is the most common modality of treatment[4].

Here, we present a 20 year old single woman who came to our outpatient department with the typical symptoms of steroid cell tumour and an uncommon symptom of ascites. She had a very dramatic response to surgical management.

Case Discussion

We present the case of a 20 year old unmarried woman who presented to us with sudden onset of amenorrhoea since 5 months. She also gives history of hirsuitism, mild abdominal distension and signs of virilisation with male pattern coarse hair growth over her chin, cheeks and anterior chest wall. She also had change in voice in the last 5 months and recent onset of dyspnoea. She had no history of feeling of mass per abdomen, loss of weight or loss of appetite. She attained her menarche at 14 years of age. Her menstrual cycles were regular initially with 3 to 4 days of bleeding every 28 to 30 days. She gave no history of any hormone intake, any other drug intake or any surgeries. She had no relevant past history, family history or personal history.

On examination, she was moderately built. She had coarse facial hair on her chin, chest and also hair on her abdomen. Her FerrimanGallaway score was 30/36. She had deep voice. She gave a history of weight gain of 6 kg over the last 5 months even though her body mass index was 24.8 kg/m^2. Her vital signs were normal with blood pressure of 110/ 70 mmHg and pulse rate of 78 beats per minute. She had no palpable mass on abdominal examination. But she had ascites with positive shifting dullness. She did not have clitoromegaly on local examination. Her secondary sexual

characteristics were normal. Per speculum and per vaginal examination could not be done as she was unmarried. Per rectal examination did not give any positive findings.

Ultrasound of the abdomen showed that uterus was anteverted 5.9* 3.3 cm with ET– 7 mm, right ovary showedhyper echoic mass lesion with centralhypo echoic component (6.6* 5.3 cm) with moderate degree of vascularity. Moderate ascites seen. Resistive index was 0.39. Left ovary was normal with no other pelvic lesions. MRI pelvis showed well defined lobulated soft tissue mass density lesion in the pelvis with non-enhancing cystic areas.

With these findings, a functional tumour of the ovary was suspected. With her age in mind, beta hCG was sent along with alpha fetoprotein, CA-125, and serum testosterone. The values are given in the table below.

	CA 125	ß HCG	Alpha FP	LDH	s. testosterone
normal	< 35	<30mIU/ml	.5-5.5 IU/ ml	140-280 U/l	0.1-10 ngm/ml
patient's values	38.14	< 1	0.35 IU/ml	214	167.1

Table 1: Various hormonal parameters in this patient as compared to normal

Peritoneal fluid analysis was done which showed no malignant cells. All the hormonal profile being normal with only serum testosterone high and no signs of malignancy patient underwent right sapling oophorectomy. Intraoperatively, the left ovary and tube was normal.

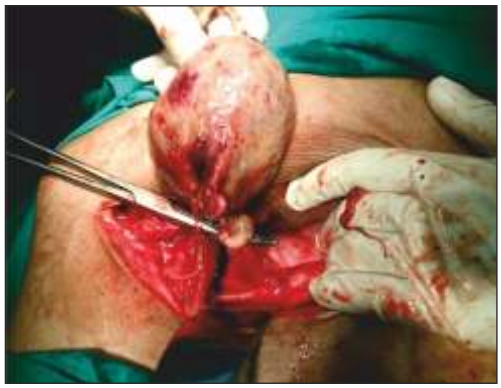

Figure 1 : Operative picture of left saplingoophorectomy

The specimen morphologically is a 6× 7 cm firm, tan brown well-encapsulated mass.

Figure 1 : morphological specimen tan brown well encapsulated mass

Figure 2 : Cut section shows haemorrhagic areas

The specimen was sent for histopathology evaluation. Histopathology relieved diffuse sheets of large polygonal tumor cells with vacuolated cytoplasm and vesicular nuclei along with nuclear pleomorphism.Crystals of Reinke, which are usually seen in hilus tumors, were not seen.

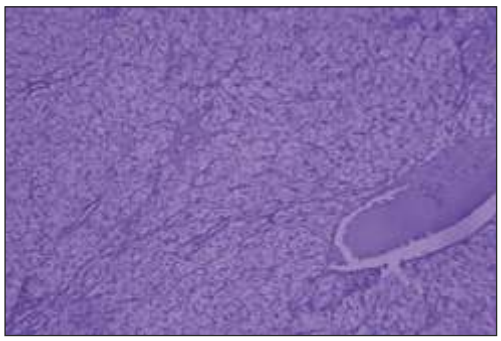

Figure 2 : diffuse sheets of large polygonal cells with vacuolated cytoplasm and vesicular nuclei and nuclear pleomorphism

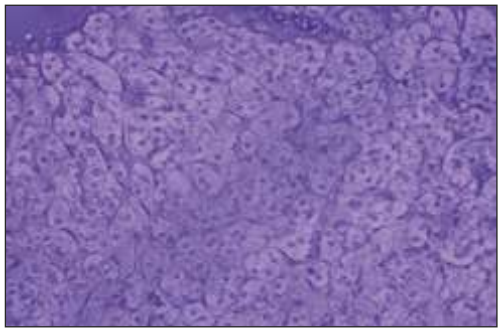

Figure 3 : magnified image of steroid cell tumour.

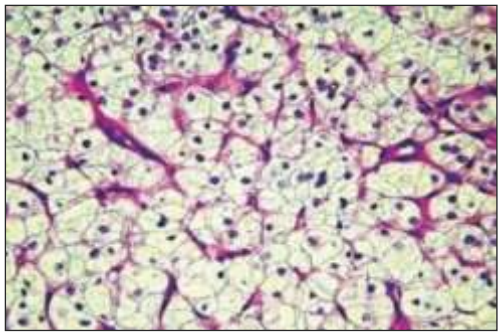

Figure 3 : Large polygonal cells with vacuolated cytoplasm and pleomorphic nucleus.

6 weeks post laparotomy, her testosterone levels attained normal values. Hirsuitism and signs of virilization was reverted and menstruation attained 2 months later. She is being called for follow up every 6 months. She is getting regular menstrual cycles with normal ultrasound abdomen and normal testosterone levels 6 months post oophorectomy.

Discussion

In 1979 Scully coined the term “steroid cell tumour of the ovary”. These tumours were previously called as “lipid cell or lipoid cell tumours”, “adrenal cell rest tumours”, “masculinovoblastomas”, “leutomas”, “hypernephroid tumours” etc as the cell of origin was controversial[1].

Ovarian steroid cell tumours account for less than 0.1% of all ovarian tumors. They are grouped under sex-chord stromal tumours and are usually benign, unilateral and characterized by a steroid cell proliferation. The name describes the morphological appearance and the clinical manifestations of the tumour. They are again divided into 3 types: not otherwise specified, stromal luteomas and leydig cell tumours. NOS subtype includes two types of polygonalcells that differ only in their cytoplasmic appearance(eosinophilia vs vacuolated). In a series of cases fromMassachusetts General Hospital, 94% of the tumors werefound to be unilateral and 28.6% were malignant[2].

The clinical manifestations of the tumor are associated with its hormonal activity and virilising properties. Among the patients affected with the tumors, 56-77% have virilising symptoms, including hirsutism, acne, clitoral enlargement, a deep voice and alopecia, and 6-23% have estrogenic manifestations, such as menorrhagia, postmenopausal bleeding or even endometrial cancer. Cushing's syndrome occurs in only 6-10% of the cases, while gynaecological examinations or surgery reveal that 25% are not associated with hormonal disturbances[7,8,9,10,11,12]

Pathological examination is an important method to diagnose steroid cell tumors. The gross specimen shows a tumor with a clear boundary, almost always well circumscribed and solid, with an enveloped, lobulated or nodular appearance[13,14,15.]

The average diameter of the tumor is 8.5 cm, and the cut surface is yellow or orange, with occasional bleeding or cystic degeneration. Microscopically, the tumor cells are round or polygonal, and medium-to-large in size with distinct cell borders. The tumor cells often have central nuclei and prominent nucleoli[14].In contrast to Leydig cells, the clear cells lack crystals of Reinke in the cytoplasm [15]

Steroid cell tumors are generally composed of granular eosinophilic or vacuolated cytoplasm which is often positive for fat stains. In addition to these microscopic findings, steroid cell tumors would require immunohistochemical markers for the accurate diagnosis. Inhibin and calretinin are the most useful markers for the discrimination of sex cord stromal tumors from other tumours. And sex cord stromal tumors are mostly negative to EMA[16].

Although steroid cell tumors are generally benign, there is a risk of malignant transformation and clinical malignant formation[1.]

The differential diagnosis of steroid cell tumor NOS includes stromal luteomas, Leydig cell tumors, luteinized thecomas, luteinized granulosa cell tumors, pregnancy luteomas and carcinomas, primary clear cell carcinomas, metastatic renal cell carcinomas and adrenocortical carcinomas. All these were excluded prior to reaching the diagnosis of a steroid cell tumor NOS.

Surgery is the most important and hallmark treatment, and complete excision of the tumor could provide the regression of symptoms and disappearance of the virilizing effects. We performed laparotomy because of the risk of malignancy and the risk of rupturing the cyst while performing laparoscopy. We performed right sapling oophorectomy since there was no evidence of malignancy.

The first-line treatment is surgery. In older women, total abdominal hysterectomy and bilateral sapling oophorectomy are the appropriate management options, while in young women, unilateral sapling oophorectomy is adequate in most cases if histology shows no malignant features. Regular follow-up evaluation with measurement of serum testosterone level is mandatory[5].Additionally a gonadotropin-releasing hormone agonist could be used as postoperative adjuvant therapy. Patients with large ovarian tumors and with an advanced stage have worse prognosis.

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