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Sertoli leydig cell tumors of ovary

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Introduction

Sertoli leydig cell tumors are the sex cord stromal tumors of ovary, are extremely rare; and account for 0.2-0.5% of all ovarian cancers[1-3]. These tumors may present with multitude of gross appearance depending upon histological grade [3].

Case report

A 16 year old unmarried girl was referred to our outpatient department on 12th February 2002 with pelvic sonography report of right ovarian mass. Her chief complaints were absence of periods, pain in lower abdomen, abnormal distribution of hair and change of voice during the last 6 months. Patient achieved menarche at the age of 12 years. She had regular cycles for 1 year which became irregular coming at 40-60 days interval for 2 years thereafter. She had amenorrhea since 6 months.

General clinical examination showed typical features of hirsutism in the form of abnormal distributions of thick, coarse hair on chin, sides of cheeks both limbs and abdomen. She had virilization as seen by clitoromegaly (Figure 1), male pattern of pubic hair distribution, and breast atrophy. Abdominal examination showed a single well defined, smooth surfaced, mobile, nontender, firm mass of about 8-10cms size in right iliac fossa. Rectal examination confirmed the findings and showed a normal sized firm, uterus.

Her routine hemogram and chest x-ray were normal. Pelvis sonography was suggestive of a right ovarian mass of 91x69mm size with internal echoes and normal sized uterus with empty cavity. Serum testosterone levels were raised to 2.41 ng/mL. Normal value in premenopausal women - 0.15 to 1.10 ng/mL).

With a clinical diagnosis of right ovarian stromal tumor a laparotomy was done on 19th February 2002. Laparotomy showed no ascitis, a single mobile, well-encapsulated, solid right ovarian mass of 10x10cm size and normal uterus and left ovary. Peritoneal fluid from multiple sites was collected for cytology and right salpingo oophorectomy done. After frozen section report of sertoli leydig cell tumor multiple peritoneal biopsies and omental biopsy were taken. Postoperative period was uneventful. Histology of the ovary reported a poorly differentiated sertoli leydig cell tumor. She received adjuvant chemotherapy of BEP Bleomycin 20 U/m^2 IV x 3 weeks, Etoposide 100 mg/m^2 IV on days 1 to 5 every 3 weeks and Cisplatin 20 mg/m^2 IV on days 1 to

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5 every three weeks regime for 3 cycles with oral contraceptives. She was relieved of symptoms after 6 months of treatment. Hirsutism and clitoromegaly disappeared within 2 months and her menstrual cycles became normal after 6 months. Her serum testosterone level came down to normal after first cycle of chemotherapy. She is having regular follow up every 2 months. At the last follow up in March 2005 (3 years after surgery) she was disease free.

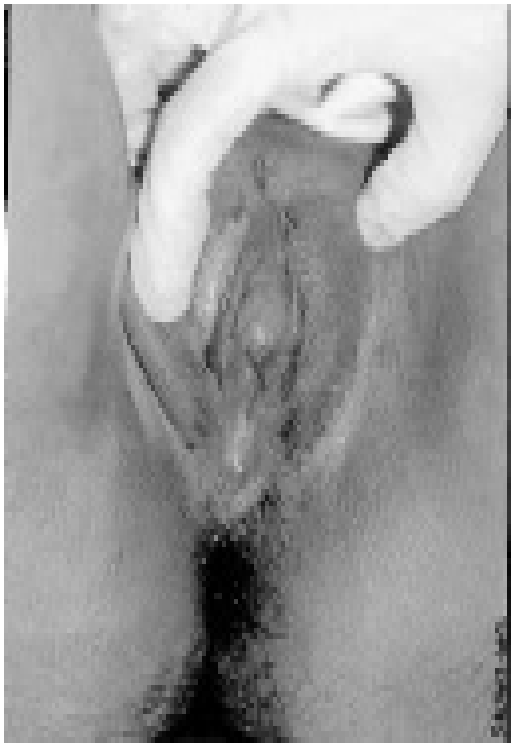

Figure 1. Clitoral hypertrophy.

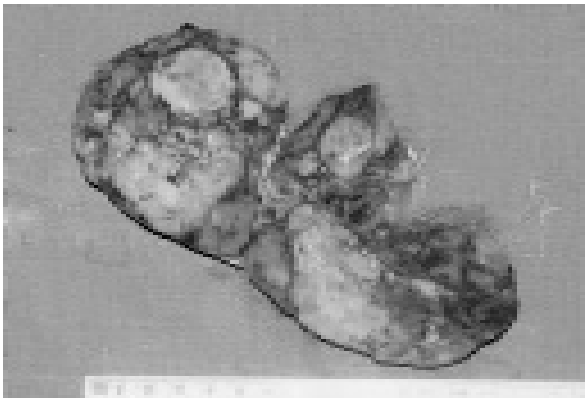

Figure 2. The ovarian mass cut open

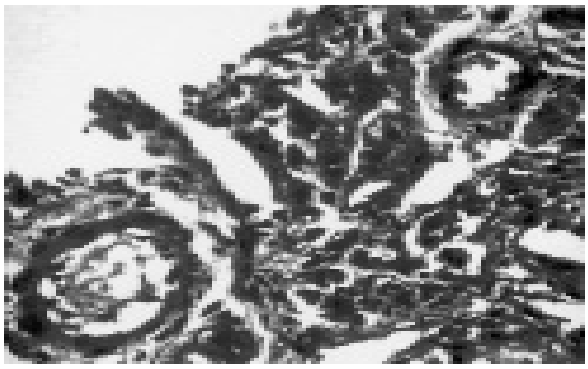

Figure 3. Clusters of Leydig cells and spindle cells around tubules (H&E staining 20x).

Discussion

Sertoli leydig cell tumors also called androblastoma or arrhenoblastoma (arrhen-male) are characterized by presence of testicular structure [2]. In 97% of cases they are diagnosed in stage Ia. In 98% of cases they are unilateral at the time of diagnosis [1-3]. The most frequent complaints of these young healthy adults at the time of presentation are menstrual disorders, virilization, and nonspecific symptoms resulting from an abdominal mass. These tumors most frequently occur in 3rd and 4th decade (85%) and in 10% of cases occur at both extremes of age [1]. Stromal tumors are well differentiated in 10%, intermediate and poorly differentiated in 50-60% and with presence of heterologous elements and retiform pattern in 30% [1]. With the exception of well differentiated tumors all have high malignant potential [2]. About size of the tumors, well differentiated ones are 5cm in size and intermediate and poorly differentiate ones > 15cm in size [3]. Familial occurrence and associated thyroid abnormalities have also been reported [2].

These tumors are hormonally active in 85% of cases and produce excess androgens mainly testosterone responsible for signs of progressive defeminization – oligomenorrhea, atropy of genital and breast tissue, and of virilization - clitoromegaly, hirsutism, increased libido, acne, hoarseness of voice, temporal balding [2,3]. A few produce estrogen from leydig cell component of the tumor resulting in estrogenisation - isosexual precosity, and irregular or postmenopausal bleeding, occasionally associated with endometrial carcinoma [3-4]. During reproductive years defeminization usually precedes virilization[2]. Other unique secretary products are serum α-feto-proteins and serum inhibin which are raised in some cases from and are Leydig cell component only [2,3]. Oliva et al [5] report a study of 54

cases including immunohistochemical profile of 23 cases. They emphasize the importance of immunohistochemistry in excluding tumors like endometrioid carcinoma and carcinoid tumor mimicking sertoli cell tumors.

Management is individualized. Staging laparotomy and complete surgical excision remain the cornerstone of treatment [6]. In stage Ia disease in younger patients fertility preserving surgery of unilateral salpingo oophorectomy with frozen section analysis, followed by staging procedure is indicated. Chen et al [7] state that no standard therapy exists. But since majority of tumors are unilateral in young patients conservative surgery is adequate while poorly differentiated tumors with mesenchymal heterogeneity need adjuvant chemotherapy. In elderly patients past reproductive age total abdominal hysterectomy with bilateral salpingo oophorectomy and infracolic omentectomy is indicated and in advanced cases additional cytoreductive surgery is required [2,3,5].

Poor prognosis is associated with extraovarian spread at the time of diagnosis, intermediate and poor grade tumors, and presence of reticular pattern and heterologous elements [2-6]. Recurrence is unusual in well differentiated tumors but in poorly differentiated type 20% have recurrence within 12 months of initial treatment. Most patients with recurrence die within 2 years [2,4,6]. Stage Ia and well differentiated tumors can be observed; all others require adjuvant chemotherapy with VAC, (Vincristine, Actinomycin D, Cyclophosphamide) PACT, (Cisplatin, Adriamycin Cyclophosphamide) BEP regime [2,3].

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