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Case Report

OVARIAN SERTOLI LEYDIG CELL TUMOUR: A RARE CASE OF SEX CORD STROMAL TUMOUR

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Abstract
Sertoli Leydig cell tumor (SLCT) of ovary is an unusual neoplasm that belongs to a group of Sex Cord-Stromal tumors of Ovary and account for less than 0.5%of all primary ovarian neoplasms. Very few cases have been documented in the literature so far. Herein we report a case of Primary Sertoli-Leydig cell Tumor (SLCT) in a 16-years old girl who presented with amenorrhea of 7 months along with hirsutism and hoarseness of voice.

Introduction

A Sertoli cell tumor (SLCT), is a Sex cord-gonadal stromal tumor of a Sertoli cells. Although Sertoli cells normally occur only in the testis, this type of tumor may also rarely occur in the ovary of females. [1] Most tumors are unilateral, confined to the ovaries, and are seen during the second and third decades of life. These tumors are characterized by the presence of testicular structures that produce androgens. Hence, many patients have symptoms of virilization depending on the quantity of androgen production. The second characteristic feature of these tumors is the degree of differentiation of structures in them. The presence of these structures determines whether the tumors are benign or malignant [2]. Twenty percent of SLCTs exhibit heterologous elements represented by endodermal elements such as cysts and glands and mesenchymal elements such as bone, cartilage or skeletal muscle. A gastrointestinal structure is rarely reported in these tumors.[3,4]

Clinical signs and symptoms can be related to either Hormonal production (mostly androgen and rarely estrogen) or presence of mass – occupying lesion (mostly pelvic-abdominal mass and / or pain).Clinically patient presents with signs of defeminization followed by masculinization. There is Breast atrophy; oligomenorrhoea followed by amenorrhoea, deepening of the voice, hirsutism, a male pattern of hair growth and clitoromegaly.

Elevated serum levels of testosterone and androstenedione can be identified in approximately 80% of patients with Ovarian SLCTS and virilizing manifestations. A

conclusive diagnosis is usually made via histology, as part of a pathology report made during or after surgery.

The usual treatment is surgery. The surgery usually is a fertility-sparing unilateral salpingo-oophorectomy. For malignant tumors, the surgery may be radical and usually is followed by adjuvant chemotherapy, sometimes by radiation therapy. In all cases, initial treatment is followed by surveillance, because in many cases Sertoli–Leydig cell tumor does not produce elevated tumor markers [5]

Herein we report a case of Primary sertoli-Leydig cell Tumor (SLCT)in a 16-year-old girl who presented with amenorrhoea of 7 months along with hirsutism and hoarseness of voice along with its literature review.

Case Report

A 16-year-old unmarried girl of Islamic religion presented to Gynecologic outpatient clinic with complaints of amenorrhoea of 7 months duration. Further history was elicited by leading and directed questions as she has not realized this symptom as a major issue. There was a history of gradual change in voice in the form of hoarseness since 7 months associated with gradual weight gain .She also C/O male pattern hair growth since last 3 months. No H/O Breast regression, anorexia ,shortness of breath, increased libido or constipation. Past medical and surgical histories were unremarkable.

Her general physical examination including breast examination was normal except for the presence of Hirsutism and Hoarseness of voice. Other vital data was normal as per her age.

An Ultrasound examination of the pelvis showed a 4.8x4.0 cm well-defined homogenous solid lesion with mildly increased vascularity in right Ovary. The left Ovary and Uterus were normal. There was no evidence of ascites. Other abdominal organs were normal.

A Hormonal profile in blood showed Increased total serum levels of testosterone of 2.27 ng/ml (normal 0.2 – 0.7) however levels of CA –125, CEA was normal.

Chest radiograph showed no evidence of pulmonary nodules.

Laparoscopic Cystectomy was planned for the patient. Operative findings showed no ascites,1.5 inches well defined soft tumor mass with a smooth surface was identified. Cyst with intact capsule without spill was removed with $1/10^{th}$ healthy ovarian tissue remaining. There was no spillage of tumor cells during surgery. The abdominal cavity was explored systematically but there were no deposits anywhere else in the cavity.

Right, ovarian cystectomy was performed with peritoneal biopsies from the right and left Paracolic gutters and Omentum. Left tube and Ovary with Uterus was found to be apparently normal and preserved. Left Ovarian biopsy and Lymph node dissection were not done.

A pathological specimen of ovary showed multiple irregular grey-brown and friable tissues together measuring 8 x8.2x2cmwith one fragment showing cyst wall measuring 3.5cm in length. Wall thickness ranges from 0.2cm to 0.3 cm.

Histopathological Examination showed a tumor composed of cords, sheets, and nests of Sertoli-like cells (hyperchromatic nuclei with moderate clear vacuolated cytoplasm). Interspersed between these are nests of polygonal cells with round nuclei and abundant granular cytoplasm (Leydig cells).

Figure 1

Mitotic activity was sparse. Sections from Right and Left Paracolic gutter and Omental Biopsy were free of tumor.

Based on the above findings , a final diagnosis of Ovarian Sex Cord tumor (SertoliLeydig Cell) Meyer Type II intermediate grade was made.

Postoperative period was uneventful and the patient was discharged on the 2nd postoperative day.

The patient was sent to medical Oncologist for further follow up .Blood tests were performed. Her serum Inhibin level was 100.89pg/ml (normal 22-85pg/ml)which was slightly raised. All other blood tests like Alpha-fetoprotein ,beta hCG, DHEA, Serum Estradiol were within normal limits.

No chemotherapy was decided for the patient but monthly follow-up of Serum Inhibin and serum testosterone is being done to rule out recurrence.

The patient resumed menses one-month post surgery and hoarseness of voice has decreased. For facial hair, the patient had taken treatment from a dermatologist.

Discussion

Sex cord-stromal tumors are groups of tumors composed of granulosa cells, theca cells, sertoli cells, leydig cells and fibroblasts of stromal origin, singly or in various combinations. They account for approximately 5-8% of all ovarian tumors and because they are rare tumors, there is a limitation in understanding their natural history, management, and prognosis. Women older than 50 years of age show the most incidences of ovarian sex cord-stromal tumors although a significant proportion occurs in premenopausal women. Leydig Stromal cell tumor is a rare case of these tumors that mostly occurs in postmenopausal women [6]. While sertolileydig cell tumors are rare and only account for ~0.5% of all ovarian tumors and can present at any age, they typically present <30 years old, with a mean age of 14 years.[7] Patient in the case reported here was of age 16 years which strengthens the case reported earlier.

Ovarian tumors that induce virilization (like hirsutism, enlargements of the clitoris, the voice deepening, etc) constitute less than 0.2% of hyperandrogenic cases. The time to onset of symptoms is usually faster in these tumors than the other causes of hyperandrogenism such as polycystic ovary syndrome (PCOS) [8], similar to our patient whose symptoms appear within months.

Meyer described three histological tumor types based on tumor differentiation. Type 1-well differentiated tubular adenoma composed of uniform tubules and mature sertoli cells. Type 2: Intermediate type composed of testicular tubules in all stages of gonadogenesis. Type 3: Undifferentiated sarcomatoid type with pleomorphic stroma and rudimentary tubule formation. WHO added Type 4 to the classification in which the tumor contains heterologous elements .In our case, it was Meyer Type 2 classification (15).

Immunohistochemistry is useful in distinguishing SLCT from other tumors. The immunocytochemical characterization of SLTC reveals positive stains of the Sertoli and Leydig cells for testosterone and estradiol. Testosterone synthesis takes place predominantly in the tumor Leydig cells, but also to a small extent in the tumor Sertoli cells [9]. The areas with tumor Sertoli cells are positive for keratins and vimentin, and negative for epithelial membrane antigen (EMA), placenta-like alkaline phosphatase (PLAP), carcinoembryonic antigen (CEA), CA 19.9, CA 125 or S-100 protein [10]. The association of positive staining for alpha-inhibin and negative staining for EMA supports the diagnosis of a stromal sex cords tumor [11]. These results show that the inhibin immunostaining may be useful in the differential diagnosis, but inhibin negativity does not exclude a diagnosis of sex cord tumor [12].In the reported case serum Inhibin level was 100.89pg/ml (normal 22--85)which was slightly raised.

Imaging features are nonspecific and variable. SLCT may manifest as a well-defined, enhancing solid mass [13] or as a cystic lesion. It typically presents as a

solid mass with intratumoral cysts.[14] Same are the finding in the present case.

After excision of the tumor ,there is a regression of hirsutism and refeminization of body configuration. Clitoral enlargement and deepened voice usually persist or show very little change. The feminization is the result of the absence of androgenicity rather than a positive estrogen effect. Patients should be followed up on serum Testosterone levels every 3 months during the first year, every 4 months during the second year ,every 6 months during the third year and thereafter annually for rest of life. As most recurrences occur within 36 months but are known to occur as late as 35 years ,life-long follows up is mandatory(16)

Conclusion

SertoliLeydig cell tumor is a rare gonadal tumor of sex cord stromal type. Clinically ,patients present with signs of defeminization followed by masculinization. It should be kept in mind as a differential diagnosis in a young female presenting with the complaints of amenorrhoea, hirsutism, and hoarseness of voice and through investigations should be done accordingly.

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Figure 1.Histopathological presentation of SertoliLeydig Cell Tumor