CASE REPORT

Sertoli-Leydig Cell -A Rare Male Hormone Producing Ovarian Tumor

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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. It accounts for less than 0.5% of all primary ovarian neoplasms. We report a case of primary intermediate ovarian Sertoli-Leydig cell tumor (SLCT) involving the left ovary in a 32-year-old nulliparous woman who presented with history of secondary amenorrhea for 8 months, hirsutism, and voice changes.
Key words: Sertoli-leydig cell tumor, ovarian tumor, secondary amenorrhea.

INTRODUCTION:

Sertoli-Leydig cell tumor (SLCT) is a rare ovarian tumor that belongs to the group of sex-cord stromal tumors. These constitute less than 0.5% of ovarian tumors.Most tumors are seen during the second and third decades of life[1]. 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] Most of these tumors are unilateral and diagnosed in stage I, so conservative surgery in a young patient is an appropriate treatment. There have also been case reports of successful laparoscopic management of the tumors.Adjuvant chemotherapy is considered for patients who have poor prognostic factors.[3]

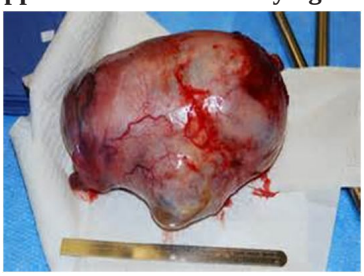
We present the case report of a young woman with Sertoli-leydig cell tumor of intermediate type limited to only one ovary.

CASE REPORT:

A 32-year-old nulliparous woman presented in Gynaecology outpatient department, with complain of secondary amenorrhea of 8 months duration preceded by menstrual irregularity (Oligomenorrhea) for last 1 year. She had also noticed a change in her voice and hair growth on her face. She was married for the last 2.5 years and had history of primary infertility. She denied any history of anorexia, weight loss, increased libido, or breast recession. Her medical and family history was unremarkable.

Her general physical examination was normal. Her height was 5 feet 4 inches and weight was 66 kg. Perineal inspection revealed mild clitoral hypertrophy. Vaginal examination revealed a firm and mobile mass of 6x6 cm in the left adnexa. Transvaginal ultrasound revealed a 5cm by 6cm solid cystic mass in the left ovary. The right ovary and the uterus were normal. Both adrenal glands were also normal. MRI confirmed the findings of ultrasound with no metastatic deposits. Her thyroid profile, S.FSH levels were normal but raised S.LH. A hormonal profile in blood indicated excessive androgenic activity in the form of elevated serum testosterone level (475.5 mg/dL; normal, 8.4-48.1 mg/dl), serum androstenedione (6.22 mg/dl: normal, 0.3-3.3mg/dl), serum free androgen index (56.6%; normal, 0.51- 6.53%). However, levels of sex hormone binding globulin, dehydroepiandrosterone sulfate (DHEAS), CA 125, and alpha-fetoprotein (AFP) were normal. On the basis of these findings, a provisional diagnosis of androgen-producing ovarian tumor was made. Exploratory laparotomy was performed. Operative findings showed replacement of left ovary by a 5 x 6-cm solid cystic, grey-white, smooth-surface intact mass. No deposits were found in the abdominal cavity and para-aortic lymph nodes were not enlarged. The right ovary was normal. Left oophrectomy was done. Peritoneal washings were sent for cytological examination. An omental biopsy was also taken. However, lymph node dissection was not done. Patient had an uneventful recovery. The histopathology report showed an intermediate (borderline) type of sertoli-leydig cell tumor. Immunohistochemical staining showed that tumor was positive for inhibin and cytokeratin. Peritoneal washing did not reveal any abnormal cells. No chemotherapy was done. During 6 months of follow-up, she started having normal periods, with resolution of her virilization symptoms. Repeat testosterone levels on follow-up were within normal range.

Fig: 1
Gross appearance of Sertoli-Leydig cell tumor



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DISCUSSION:

Sertoli-Leydig cell tumors are rare of all ovarian tumors, as in one study of Pakistan [4] only 7% of sex cord stromal tumors were SLCT. While they can be found in women of all age groups, they are most common in young women. Our patient presented at young age in contrast to two studies that reported a teenage girl and a postmenopausal woman.[5,6] Virilization in females can occur based on ovarian or adrenal pathology. In terms of ovarian-based female virilization, Sertoli-Leydig cell ovarian tumors produce and release a male sex hormone which may cause the development of male physical characteristics including facial hair and a deep voice[7]. Similar features were present in this case also.

In our patient, ovarian tumor was diagnosed on ultrasound. Hormone assessment was done due to development of male hormone symptoms.In the surgical treatment of SLCTs it is necessary to adopt common guidelines, and evenly define the steps that the patient should be submitted. Laparotomy was done in this patient; howeverthere have also been case reports of successful laparoscopic management of these tumors.[3] Different steps that are usually used for oncological ovarian cancer staging are not always performed. Conservative and fertility sparing surgery is commonly accepted, and even preferred due to the young age of patients as done in our patient and another study.[8] The important prognostic factors in these tumors are their stage and degree of differentiation. In a review of 207 cases by Young and Scully in 1985 1 all well-differentiated tumors were benign, whereas 11% of tumors with intermediate differentiation, 59% of tumors with poor differentiation, and 19% of those with heterologous elements were malignant. In the present case report histopathology report showed intermediate differentiation which has a good prognosis. Of various immunohistochemical stains applied, Inhibin and cytokeratins were positive in the histopathology report of this patient SLCT, as same results were found in another study.[4]

In this patient as tumor was of early stage and of intermediate type so chemotherapy was not recommendedin contrast to a study [9] whereincomplete staging of the tumor and the presence of heterologous elements on histopathology favors for chemotherapy. The malignancy rate in tumors with heterologous elements is 15% to 20%. Adjuvant chemotherapy in stage I is given to those patients who have poorly differentiated SLCT or SLCT with heterologous elements or a metastatic tumor of any histologic type. The BEP (Bleomycin, Etoposide, Cisplatin) regimen is a comparatively safe chemotherapeutic regimen because it does not affect the fertility status of the patient.[10]During her 6 months follow up, patient had resumed normal menstrual cycles and her voice also became feminine as another study concluded that feminine characteristics return after surgery, but manifestations of masculinization disappear more slowly. [9]

In conclusion, SLCT is a rare ovarian sex-cord tumor that usually occurs unilaterally. SLCT should always be considered in a young female patient who has symptoms of virilization and an ovarian mass on examination or investigation.Management depends on the histopathology of the tumor. Intermediately differentiated tumors need an individualized approach.

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