

Androblastoma of the Ovary

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Androblastoma of the ovary occurs most frequently in the 3rd to 4th decades of life and about 75% of the lesions are seen in women younger than 40 years. This tumor produces androgen and virilization is present in more than 70% of the patients. Due to the early diagnosis and treatment of such type of tumor, the prognosis is very good. This was a case report of an 18 years' unmarried girl who presented with signs of virilization and amenorrhea for two years. On abdominal examination mass was up to epigastric region, after all investigation we planned for laparotomy. Histopathology report revealed androblastoma of the ovary. After a month of operation, she had regular menstruation.

Keywords: amenorrhoea; androblastoma; breast atrophy; deepening of voice; hirsutism.

INTRODUCTION

Androblastoma is a Sertoli Leydig cell tumor (SLCT) of the ovary. It has low malignant potential. It is extremely rare tumor and accounts for only less than 0.2% among all ovarian tumors. It is most commonly found in the third and fourth decades of life and 75% of the lesions are seen in women younger than 40 years.¹ This tumor produces androgen, hence the signs of virilization like oligomenorrhea followed by amenorrhea, atrophy of the breast, acne, hirsutism, cliteromegaly and deepening of the voice are commonly seen.² The level of male hormones like plasma testosterone, androstenedione and DHEA-S are high. The diagnosis of androblastoma is confirmed by histopathology reports.¹

CASE

An 18 years unmarried girl was referred to our gynaecology outpatient department with a complaint of amenorrhea for two years, abnormal distribution of hair since same duration, hoarseness of voice since one year and gradual distension of abdomen since six months, which occupied the whole abdomen. She did not have a history of loss of weight and appetite.

There was no family history of gynaecological malignancy. On examination, her general condition was good but she had hair growth all over the body. Per abdominal examination, abdomen was grossly distended up to epigastric region, cystic in consistency with fullness in bilateral flanks; surface was smooth with ill-defined margin but non-tender. Pelvic examination revealed huge mass. Uterus was not felt separately and the origin of mass could not be identified separately as the mass was bigger in size. Ultrasonography revealed large, mixed echogenic mass predominantly anechoic lesion noted in the pelvis and extended up to epigastric region D/D right ovarian complex mass with hydronephrosis on right side. Hormone assay revealed raised serum ACTH level. CT scan of the abdomen and pelvis revealed large cystic mass measuring about 29cm x 23cm x 12cm occupying the whole abdomen and pelvis. Multiple irregular enhancing septum and minimal soft tissue components were also seen within it. No calcification and fat density were noted within the mass. Mass was displacing the bowel loop laterally and compressing the right ureter with evidence of right hydronephrosis. However, the left kidney was normal. Exploratory laparotomy findings reveals that the right ovary was about 25 cm x 20 cm x 10 cm with smooth surface, cystic consistency, intact capsule, mobile and weight of tumor was 6 Kg. The left tube and ovary was normal. Uterus, omentum, colon, liver, stomach and peritoneum were normal. No palpable lymph nodes were present. A probable surgical diagnosis was benign tumor of the right

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ovary, which could be androblastoma. Cut section revealed multiple loculation with solid cystic areas (Figure 1). Microscopically, tumor cells were arranged in lobules and were separated by the zones of loose fibrous stroma. Tumor cells were also arranged in cords irregular into the surrounding fibrous or fibromixoid stroma (Figure 2). These tumor cells have clear abundant cytoplasm with oval nuclei and conspicuous nucleoli. Nuclei were relatively bland with mitosis. There were thin walled cells lined by flattened or cuboidal cells, which create a sieve like pattern. The indifferent mesenchyme was abundant and consisted of solid cord and scanty cytoplasm. Mature leydig cells were also seen in periphery of tumor cells, which arranged in seat cluster and singly scattered. Peritoneal cytology showed absence of malignant cells. After one month of operation, she came for the follow up. She was very happy as she was having regular menstruation and her voice was also improved.



Figure 1. Cut section of tumor- well-circumscribed multilobulated mass with solid cystic area.

is heralded by hirsutism, temporal balding, deepening of voice, cliteromegaly and there may be secondary amenorrhea. Elevated level of androgen is the diagnostic clue of such type of tumor. The malignant potential is lower than epithelial ovarian tumor.¹ They are sub-classified according to the histological pattern into well differentiated grade 1; moderately differentiated grade 2; poorly differentiated grade 3; and retiform pattern (a very characteristic appearance under the microscope) tumor with heterologous elements. Fortunately, more than 97% of SLCT are grade 1.² SLCT are usually unilateral and bilateral SLCT is very rare. If it is bilateral, then there should be other pathological basis.⁵ Inhibin is a glycoprotein hormone produced by normal ovarian granulosa cells and testicular sertoli cells. Patients with granulosa cells tumor have elevated level of inhibin and which is useful in diagnosis of recurrent ovarian tumor.⁵ We estimated alpha-fetoprotein and it was high, i.e, 3.08 IU/ml (normal: 0-2 IU/ml).

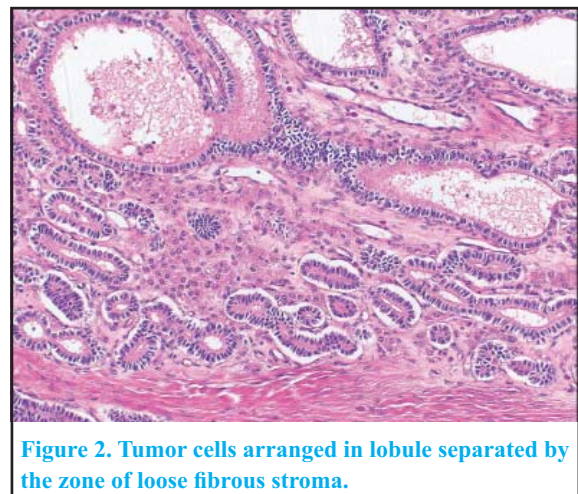


Figure 2. Tumor cells arranged in lobule separated by the zone of loose fibrous stroma.

COMMENT

Androblastoma (Sertoli-Leydig cell tumors) are rare, sex cord stromal tumors of the ovary composed of undifferentiated gonadal stromal cells, Leydig cells and Sertoli cells.³ Recent studies have shown that many cases of sertoli-Leydig cell tumors (SLCT) of the ovary are caused by germ line mutation in the DICER1 gene.⁴ SLCT occurs in all age groups. Incidence of SLCT in prepubertal age group is 5% In postmenopausal women, abnormal uterine bleeding is the initial symptom due to the high level of oestrogen. It is bilateral in 2% of cases.¹ Classically, there is progressive masculinization that

We did not estimated inhibin due to the unavailability of the assay. Serum ACTH level was also high which was 60.7pg/ml (normal: 10-46 pg/ml). Besides, other investigation such as immunohistochemistry for intermediate filament and sex hormone receptor, polymerase chain reaction (PCR) for detecting the presence of the sex-determining region in Y gene can also be done. But all of these tests are not available here.⁶ Unfortunately, the prognosis does not depend upon histological appearance of a tumor.⁷ The treatment of choice is simple-adnexectomy of the affected side with comprehensive surgical staging and follow up with regular measurement of serum androgen. The 5 years survival is 70%-90%.¹ This

patient is now asymptomatic and she is on regular follow up till date.

CONCLUSIONS

It is a very rare tumor with low-grade malignancy and very good prognosis. Any woman presented with amenorrhea with sign of masculinization should be investigated and rule out androblastoma. Treatment is

simple adnexectomy except in advance cases in which adjuvant chemotherapy is needed.

DISCLOSURE

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