Case Report

GRANULOSA CELL OVARIAN TUMOR AS CAUSE OF PRIMARY AMENORRHOEA

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ABSTRACT

An 18-year old single girl presented in gynecological outpatients department in April 2004 as a case of primary amenorrhoea and abdominal mass. Laparotomy was performed and tumor removed. Diagnosis of granulosa cell tumor was confirmed on histopathology.
Granulosa cell ovarian tumor usually presents with menstrual problem in the form of precocious puberty, irregular bleeding or postmenopausal bleeding. Our patient presented with primary amenorrhoea that is extremely rare presentation.

KEY WORDS: Primary Amenorrhoea. Abdomen. Mass. Tumor. Granulosa Cell.

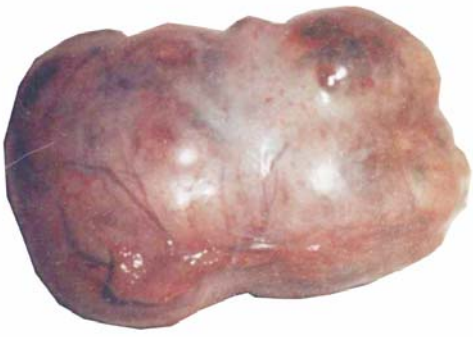
INTRODUCTION

Primary amenorrhoea is the absence of menstruation by 16 years of age in the presence of normal secondary sexual characteristics, or by 14 years of age if secondary sexual characteristics have not developed.[1] It occurs in 1-3% of women of reproductive age. Frequency of primary amenorrhoea is reported as 0.065%[2] and 0.06%[3] by two local studies. Primary amenorrhoea poses a diagnostic challenge for gynecologist that requires an understanding of normal menstrual function during pubertal years. Visible menstruation depends upon inter-related factors, which include 46 XX karyotype, intact hypothalamic pituitary ovarian axis, endometrium responsiveness to hormonal stimulation, secretion of estrogen from ovaries resulting in ovulation with the secretion of progesterone as a result of follicle stimulating harmone (FSH) and lutenizing hormone (LH) secretion and finally an intact outflow tract. The differential diagnosis is broad ranging from anatomical anomalies of the genital tract; genetic and endocrine disorders to environmental and psychological factors[4].

CASE REPORT

A young 18-year old girl presented to the outpatients department of Liaquat University Hospital, Hyderabad in April 2004 with complaints of primary amenorrhoea and abdominal mass for last 2 months. The mass had gradually increased in size. She did not give any history of pain and fever. Her bowel and urinary habits were normal. There was no history of diabetes, hypertension or genital tract malignancy in the family. On examination, she was 144 cm tall and weighed 40 kg. Her pulse was 80 beats/minute and blood pressure 110/70 mmHg. Her breasts were fully developed with no galactorrhoea; axillary and pubic hair, thyroid and external genitalia were also normal. Abdominal examination revealed a well-defined, firm, mobile, non-tender mass equivalent to 18 weeks size gestation arising from pelvis extending up to umbilicus and more towards right side of the abdomen. All base line investigations including Complete Blood Count, Liver Function Test, Renal Function Test and Midstream Urine Analysis were normal. Serum follicle stimulation hormone, lutenizing harmone, Thyroid function test, Prolactin levels were also within normal range. Buccal smears revealed female karyotyping while ultrasonography revealed anteverted normal size uterus and right adenexal solid mass of 15x10 cm size with multiple septa. Left ovary was normal and no ascites was visualized. Hence, diagnosis of right ovarian tumor was established. The girl and her parents were counseled regarding the probable nature of tumor and need for surgery and she was prepared for Laparotomy. Laparotomy findings revealed normal uterus, left ovary and fallopian tube. A 15x10cm size solid, lobulated, encapsulated mass was found arising from right ovary (**Figure I**).

FIGURE I: POSTOPERATIVE SPECIMEN OF THE TUMOR



No ascites was visualized and there was no evidence of secondaries within or outside the pelvis. A right salpingo-oophorectomy was performed and tumor was removed en-mass. Peritoneal washings were taken for cytological examination. Patient had smooth recovery with an uneventful postoperative period. Tumor histopathology report showed Granulosa Cell Tumor of Ovary with negative peritoneal washings. She was discharged on the sixth postoperative day with the advice for regular follow up. Patient came for follow up and after two months had established normal menstruation.

DISCUSSION

Granulosa cell tumor accounts for 5-10% of ovarian malignancies[5].There are two main varieties; juvenile and adult forms. Adult granulosa cell tumor is characterized histologically by Call Exner bodies, pale and grooved nuclei and the mature follicles.

Both adult and juvenile forms commonly produce estrogen, and estrogen production often is the reason for early diagnosis while clinical features are related to the reproductive status at the time of presentation. In pubertal age group, the granulosa cell tumor manifests with estrogenic symptoms such as breast enlargement, appearance of pubic and axillary hair, enlargement of external and internal genitalia and irregular uterine bleeding. In reproductive years, it presents with menstrual irregularities and in postmenopausal women with uterine bleeding as a result of endometrial hyperplasia. Well-differentiated endometrial adenocarcinoma occurs in 10-25% of cases of postmenopausal women[6]. It may occasionally rupture and results in abdominal pain, hemoperitoneum and hypotension mimicking an ectopic pregnancy. The staging system for these tumors is same as for epithelial ovarian tumor. Bilateral tumors are present in only 5% of cases[7]. Primary treatment for a patient with granulosa cell tumor is always surgical and is same as for epithelial ovarian cancers. In older woman, hysterectomy with bilateral salpingo-oophorectomy is the first option. In younger patients who desire future fertility a unilateral salpingo-oophorectomy as we done with our patient almost provide sufficient treatment because most of these tumors are stage I. For advanced stage disease more than Ia there is need for chemotherapy but as in our case, growth was confined to one ovary with intact capsule, no ascites and negative peritoneal washings, so, there was no need for adjuvant chemotherapy or radiotherapy.

Chemotherapy and radiotherapy are reserved for patients with advanced disease and for patients with recurrent tumor. In case of recurrence, further surgery should be considered before any other therapy is given for both therapeutic and diagnostic reasons. In addition to tumor stage, residual disease, patient's age, tumor size, extent of surgery[8], number of mitosis per 10 high power field or Call Exner bodies and cellular atypia have been reported to be of prognostic significance[5]. Prognosis for granulosa cell tumor generally is very favorable. Approximately 90% of granulosa cell tumors are at stage I at the time of diagnosis. The 10 -year survival rate for stage I tumor in adults is 90% treated by surgery alone[5]. Granulosa cell tumors of more advanced stages are associated with 5 years and 10 years survival rates of 33% – 44%. Granulosa cell tumors have propensity for late recurrence; some occurring as many as 37 years after detection and treatment of primary growth[5]. Mean survival after diagnosis of recurrence is 5 years. Fertility is not affected by granulosa cell tumor if detected at an early stage when growth is confined to one ovary with no evidence of extension of tumor. Because of propensity of granulosa cell tumour to recur years after initial diagnosis, prolonged surveillance with serial physical examinations and serum tumor markers such as estradiol and inhibin is reasonable[9].

CONCLUSION

To the best of our knowledge, this is the first reported case of granulosa cell tumor causing primary amenorrhoea from this part of the world. Therefore, granulosa cell tumor although rare should be considered in the differential diagnosis of adenexal mass to allow early identification, timely surgical management and excellent cure rate, good overall prognosis. Long-term follow up is always required in patients with the granulosa cell tumor.

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