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JUVENILE GRANULOSA CELL TUMOR WITH UNUSUAL PRESENTATION: A CASE REPORT

Gynaecology

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ABSTRACT

Background: Granulosa cell tumor (GCT) is an uncommon malignant neoplasm primarily arises from the sex-cord stromal cells of the ovary. They constitute only 2% of the ovarian tumors and more than 70 % of the sex cord-stromal tumors. There are two distinct histological types: Juvenile GCT represents only 5% of this tumor and usually occurs in pre-pubertal girls and women younger than 30 years whereas Adult GCTs present with early stage disease and are commonly encountered in the age group of 35 years to 55 years. These tumors have a particular clinical and histological profile, and may reoccur up to 40 years after diagnosis in 25% cases.

Case report: A 22 year old female patient, nulligravida, with 4 years of active married life, presented to gynaecology oncology clinic with lower abdominal pain, and distension and breathlessness after mild work since 1 month.. On examination, she had Pulse Rate of 96/min and her blood pressure was recorded as 110/70 mm Hg, pallor was present, on per abdominal examination, ascites was present ,mass was felt arising from pelvis extending above umbilicus and was non tender. On local examination clitoromegaly was present. Vaginal examination revealed healthy cervix and vagina and normal sized uterus and mass felt as described above in right adnexal region. All blood investigations including tumor markers germ cell tumor markers were within normal limits except for the CA-125 which was 900 U/ml and hormonal profile was normal. Ascitic fluid and pleural fluid tapping was done and approx 600 ml of pleural fluid drained out and sent for examination and showed no malignant cells in fluids. Chest radiograph showed hydro-pneumothorax and thus intercostals drainage tube was kept. With provisional diagnosis of ovarian neoplasm she was taken up for laparotomy. Final histopathological examination report showed juvenile granulosa cell tumor with intact capsule. So patient was staged 1A of ovarian neoplasm. She is on chemotherapy at present.

Conclusion: This was a rare case of juvenile granulosa cell tumor that presented with abdominal pain along with breathlessness and ascites and pleural effusion resembling germ cell type picture.

It turned out to be juvenile granulosa cell tumor with unusual presentation. Since they are less frequent and may present with unusual clinical picture, their differential cannot be overlooked in any age group.This patient carries good prognosis and 5 year survival of 95%. She is suggested for child bearing 6 months after completion of chemotherapy.

KEYWORDS

juvenile granulosa cell tumor, unusual presentation

Introduction

Granulosa cell tumor (GCT) is an uncommon malignant neoplasm primarily arises from the sex-cord stromal cells of the ovary. They constitute only 2% of the ovarian tumors and more than 70 % of the sex cord-stromal tumors.

There are two distinct histological types, Adult granulosa cell tumor and Juvenile granulosa cell tumor. Juvenile type of GCT represents only 5% of this tumor and usually occurs in pre-pubertal girls and women younger than 30 years whereas Adult GCTs present with early stage disease and are commonly encountered in the age group of 35 years to 55 years. These tumors have a particular clinical and histological profile, and may reoccur up to 40 years after diagnosis in 25% cases. [1]

They are hormone active tumors, originating from granulosa cells which produce estradiol. Overproduction of estradiol is helpful in the diagnosis of the tumor because of its numerous symptoms of hyperestrogenism. These are tumors with relatively a favourable prognosis compared to epithelial ovarian cancers. What makes them different from the epithelial ovarian cancers is the nature of presentation and clinical behaviour [2].

In view of uncommon gynaecological malignancy and to preserve the fertility, histological examination of the resected specimen always warranted with expertise. There has been a lot of interest in the molecular pathogenesis of these tumors and due to their origin from the granulose cells; they are potential targets for hormonal and targeted therapy.

Juvenile granulosa cell tumors are rare tumors, constituting only 5% of granulosa cell tumor with distinct pathological appearance. These tumors have varying clinical presentation with varying clinical course and prognosis. In view of rarity of the condition a case is summarised and discussed in the current article.

Case Report:

A 22 year old female patient, nulligravida, with 4 years of active married life, presented to gynaecology oncology clinic with lower abdominal pain, and distension and breathlessness after mild work since 1 month. She had regular menses. Her menstrual cycles were regular, with normal flow during menses.

On examination, she had Pulse Rate of 96/min and her blood pressure was recorded as 110/70 mm Hg, pallor was present, on per abdominal examination, ascites was present ,mass was felt arising from pelvis extending above umbilicus and was non tender. On local examination clitoromegaly was present. Vaginal examination revealed healthy cervix and vagina and normal sized uterus and mass felt as described above in right adnexal region.

Ultrasonography was performed which revealed a 8 x 14 x 18 cm solid lesion arsing from right adnexal region. Uterus was of normal size and ET was 7mm. Lesion showed internal cystic areas and septations. Also moderate ascites and right moderate pleural effusion was found. Tumor markers for germ cell tumors were within normal limits. CA-125 was elevated, 586 U/ml.

Ascitic fluid and pleural fluid tapping was done and approx 600 ml of pleural fluid drained out and sent for examination and showed no malignant cells in fluids. Chest radiograph showed collapse of underlying lung, necissating the need for intercostals drainage tube insertion on the right side.

With provisional diagnosis of ovarian neoplasm she was taken up for laparotomy. Intraoperatively, ascites was present (approx 400ml). Ascitic fluid was sampled for cytology studies and drained left ovary and fallopian tube was found normal. A right adnexal mass of approx

20 x 16 cm was present. Deposits over POD and omentum were found. Rest of the peritoneal cavity was normal. (fig 1)

Right adnexal mass was removed and sent for frozen section. Following the report of frozen section which showed granulose cell tumour operation was continued. Deposits over POD and bladder were biopsied. Omental sampling was done followed by right side pelvic lymph node dissection. Postoperative period was uneventful. Final histopathological examination report showed juvenile granulosa cell tumor with intact capsule. (fig 2)

The patient was planned for adjuvant chemotherapy with carboplatin and Paclitaxal. At present she has no complaints and is on treatment.

Discussion

Juvenile Granulosa cell tumors are rare neoplasm, that develop primarily in children and young adults, and approximately 90 percent are diagnosed before puberty. The mean age at diagnosis is 13 years, but patient age may range from newborn to 67 years. Juvenile granulosa cell tumors may be associated with Ollier disease or with Maffucci syndrome, which is characterized by presence of endochondromas and hemangiomas.

These tumors may be hormonaly active with elevated estrogen, progesterone, and testosterone levels. This leads to suppression of gonadotropins. As a result, menstrual irregularities or amenorrhea are common. Prepubertal girls typically display isosexual precocious puberty, which is characterized by breast enlargement and development of pubic hair, vaginal secretions, and other secondary sexual characteristics. In addition these tumors may secrete androgens, but in such cases virilization may be observed. Despite these endocrinologic signs, a delayed diagnosis of juvenile granulosa cell tumors in pre- and postpubertal girls is common. [3]

The most common presenting symptoms of both adult and juvenile granulosa cell tumors are abdominal pain and increasing abdominal girth. In 6-10% of cases, tumor rupture causing acute abdomen pain can be the presenting symptom (4).In the case that presented to us the patient has complaint of abdominal pain and distension. In addition was infertile despite 4 years of marriage without utilizing any contraceptive practices. Also patient was seen to have clitoromegaly. The patient also had ascites and pleural effusion. Exact mechanism of the development of ascites is still unknown. However a few theories relating to mismatch in arterial and venous blood flow, obstruction of lymphatics due to mechanical obstruction and pertioneal irritation secondary to the tumor have been put forth. Approximately 10 percent of the patients present with ascites. In addition to aforementioned theories, Abramov et al have put forth the role of VEGF and inflammatory cytokines expressed by granulosa cell tumor in formation of ascites. [4,5]

Juvenile granulosa cell tumors are grossly similar to the adult-type tumor and display variable solid and cystic components. They can attain significant size and have an average diameter of approximately 12 cm. microscopically, cytologic features that distinguish these tumors from the adult type are their rounded, hyperchromatic nuclei without “coffee-bean” grooves. Call-Exner bodies are rare, but often there is a theca cell component. (1) In the patient being reported a 19 cm right adnexal mass, yellow grey in colour with solid cystic areas was excised. On histopathology it was reported as Juvennile Granulosa cell tumor.

Prognosis is excellent, and the 5-year survival rate is 95 percent. Similar to adult-type tumors, 95 percent of juvenile granulosa cell tumors are unilateral and stage I at diagnosis. However, the juvenile type is more aggressive in advanced stages, and the time to relapse and death is much shorter. Recurrences typically develop within 3 years and are highly lethal. Later recurrences are unusual. The only statisitcally proven poor prognostic factor is the stafe of the tumor. In addition size of tumor, age, nuclear atypia and mitotic figures may be considered as poor prognostic markers, though not scientifically confirmed. In our patient a primary upfront surgery was performed following which the patient was diagnose to be stage IC Juvenile Granulosa cell tumor. In view of it being stage Ic with large size tumor patient was considered for adjuvant chemotherapy with carboplatin and paclitaxel.

Prolonged surveillance program with periodic clinical, serologic and radiologic follow-up is imperative, given the tendency of these tumors to recur after the initial management. [6]

Conclusion:

The case is presented due to the rarity of the tumor subtype. Juvenile granulosa cell tumor are encountered less frequently in clinical practice. Patient in this report presented with abdominal pain along with breathlessness and ascites and pleural effusion. These tumors may present with unusual clinical picture, their differential cannot be overlooked in any age group. These patients carry a good prognosis with ideal and prompt intervention with a 5 year survival of 95%.

FIGURE 1:

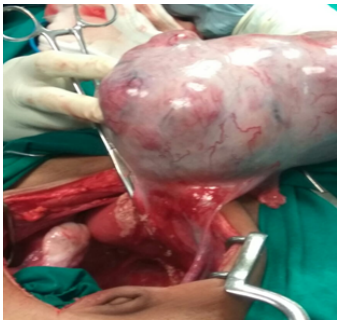
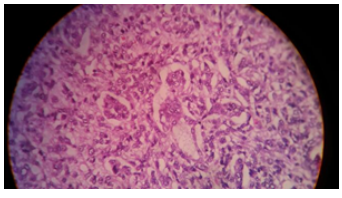


FIGURE 2:



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