JUVENILE OVARIAN GRANULOSA CELL TUMOUR: A CASE REPORT

Iornum Hembe Shambe,[1] Jamila Ali[1]

[1]Department of Obstetrics and Gynaecology, University of Jos

ABSTRACT

Granulosa cell tumours of the ovary are rare ovarian tumours but the commonest of the sex cord stromal tumours. They typically have good prognosis. Presentation in premenarchal women or those in their reproductive years is mostly managed by conservative surgery.

We present the clinical characteristics, diagnosis and treatment in an 18year old female who presented with progressive abdominal swelling and pain.

Key words: juvenile, granulosa cell, Tumour, ovary

INTRODUCTION

Granulosa cell tumours of the ovary are a rare type of sex cord stromal tumour of the ovary which typically present at early stages with good prognostic outcomes when compared to other epithelial tumours. They are usually hormonally active secreting oestrogen and are predominantly solid lesions with less common cystic, unilocular forms. They account for 1-2% of all ovarian malignancies and 95% of germ cell tumours that originate from sex cord stromal cells.[1,2]

Based on age, this type of tumour is subdivided into Adult (AGCT) and Juvenile types (JGCT) which represent 95% and 5% of cases respectively.[3] The adult types are usually common in the 5th decade of life while the juvenile types are rarely seen with a majority seen before puberty or in females aged less than 30 years. JGCTs are more likely to be hormone secreting tumours as well which may present with irregular vaginal bleeding, pseudo puberty or in virilization some rare instances. Presenting symptoms vary with the stage at diagnosis but abdominal pain and distension are common.[4]

In this case study, the clinical presentation, imaging features, as well as the histopathological features and treatment are discussed with the relevant review of literature.

CASE REPORT

A.S was an 18 year old female Po+^{o} who was referred to the gynaecological emergency unit of Jos University Teaching Hospital (JUTH) from the General Out Patient Department, with a complaint of progressive, painless abdominal swelling of 8 months duration. The swelling started in the right iliac fossa, gradually increasing in size to involve the whole abdomen. There was associated early satiety, and difficulty in breathing but no cough. There was no history of fever or change in her urinary or bowel habits. She had lost weight and had easy fatigability. She had noted a deepening of her voice since the illness started.

There was no history of abnormal vaginal bleeding, and there was no history of ovarian, endometrial or breast cancer in her family.

On examination, she was a young girl who was chronically ill-looking, pale and jaundiced. She was not dehydrated and no regional lymph nodes were enlarged.

She was tachypnoeic with a respiratory rate of 22 cycles per minute. There were vesicular breath sounds in the chest with no added sounds. The cardiovascular system was essentially normal.

The abdomen was grossly distended, with generalized tenderness but no guarding. The Intra-

abdominal organs difficult to palpate due to ascites. There was a huge intra-abdominal mass extending from the right iliac fossa to the epigastrium measuring approximately 35cm x 20cm .The mass was firm with a delineable upper limit but the lower limit could not be appreciated.

Ascites was demonstrable by a fluid thrill

A pelvic examination showed clitoral hypertrophy with an intact hymenal ring intact .There was good perianal hygiene. A digital rectal examination showed good sphincteric tone, the rectal mucosa was freely mobile with fullness felt at anterior rectal wall.

An impression of an ovarian Tumour was made.

INVESTIGATIONS: Her Packed Cell Volume (PCV) was 20% and a White blood cell count showed a lymphocytosis of 14.6 x 10^9 cells accounted for by a neutrophilia of 83%.The urea and electrolytes and Liver function tests were essentially normal.

Abdomino-pelvic Scan showed a huge right adnexal mass with cystic and solid components. An abdominal CT scan showed a huge Intra-abdominal complex cystic mass.

The Bed site clotting time was 6 minutes

Alpha-fetoprotein (AFP) was elevated - 343.5ng/ml, Lactate dehydrogenase was also elevated LDH – 413.0Iu/l while CA 125 levels were normal. The Hepatitis B test and Human immunodeficiency virus assays were non-reactive.

She was transfused with 3 units of blood over 3 days and subsequently had staging laparotomy where a Right total salpingo-oophorectomy and an infracolic omentectomy was performed with the following findings.

- 4 litres of straw coloured ascitic fluid + 1 litre of bloody fluid from a ruptured cyst which also contained some yellowish green gelatinous substance
- Huge Right Ovarian trilobular masses with the largest measuring about 30cm x 22cm x 10cm.Capsule intact
- Grossly normal left tube and ovary with normal sized healthy looking uterus
- Grossly normal liver and spleen. No tumour seedlings, No intra-abdominal nodules palpated.
- Fibrous adhesions between the ovarian tumour, ileum and mesentery
- EBL – 100ml

Her post op condition was satisfactory. She was placed on Intravenous Ceftriaxone and metronidazole and given intramuscular analgesics: Pentazocine and Diclofenac .She had intravenous fluid for 48 hours.

Samples were sent for Cytology and Histology.

Cytology - showed inflammatory cells and histology showed a Granulosa cell tumour

FIGO Stage was ascertained to be IA

She was transfused with 3 more units of blood transfused after a post op PCV of 16%. She was discharged with a post transfusion PCV of 31% to the gynaecological outpatient unit. When reviewed in the clinic 3 weeks post operatively, the deepened nature of her voice had reduced and the clitoral hypertrophy observed before surgery, had resolved.

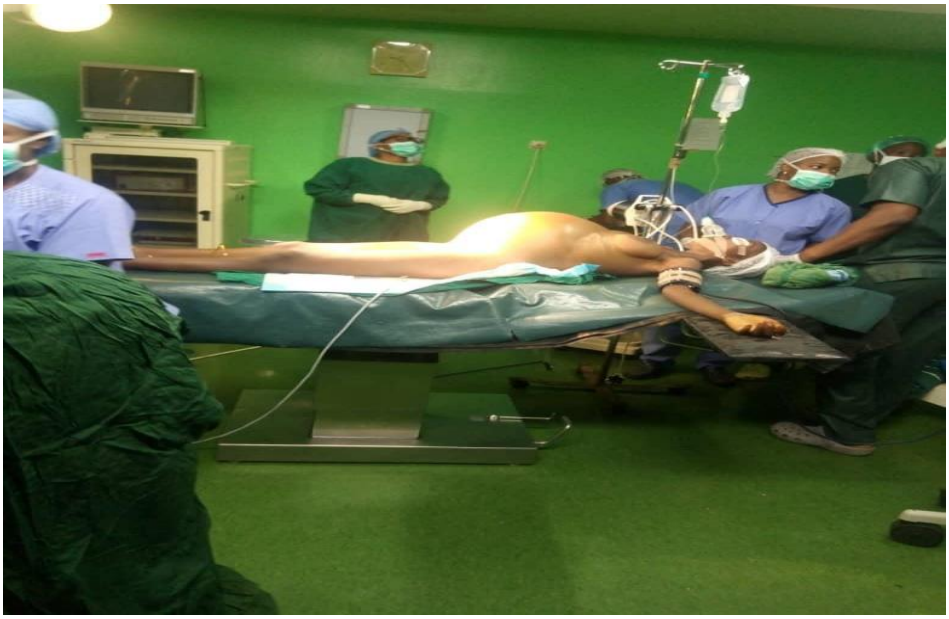

Figure 1: Miss A.S after being anaesthetized shortly before surgery

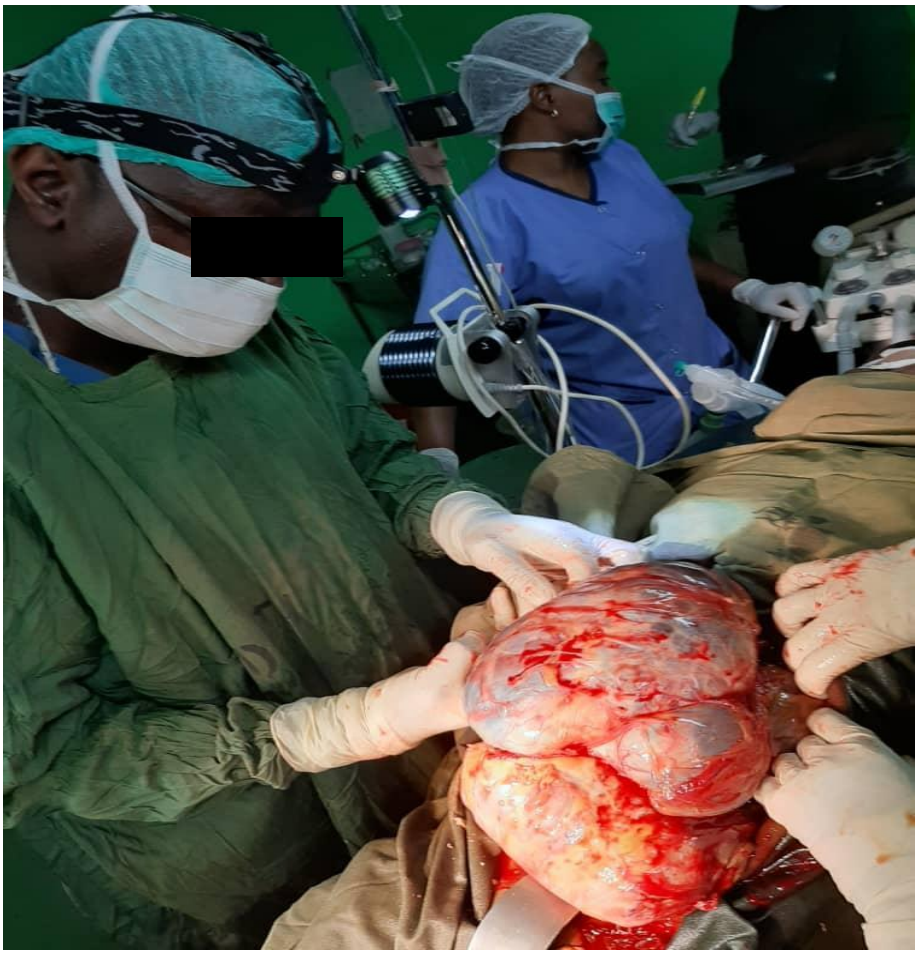

Figure 2: The ovarian tumour after being mobilized from the abdominal cavity

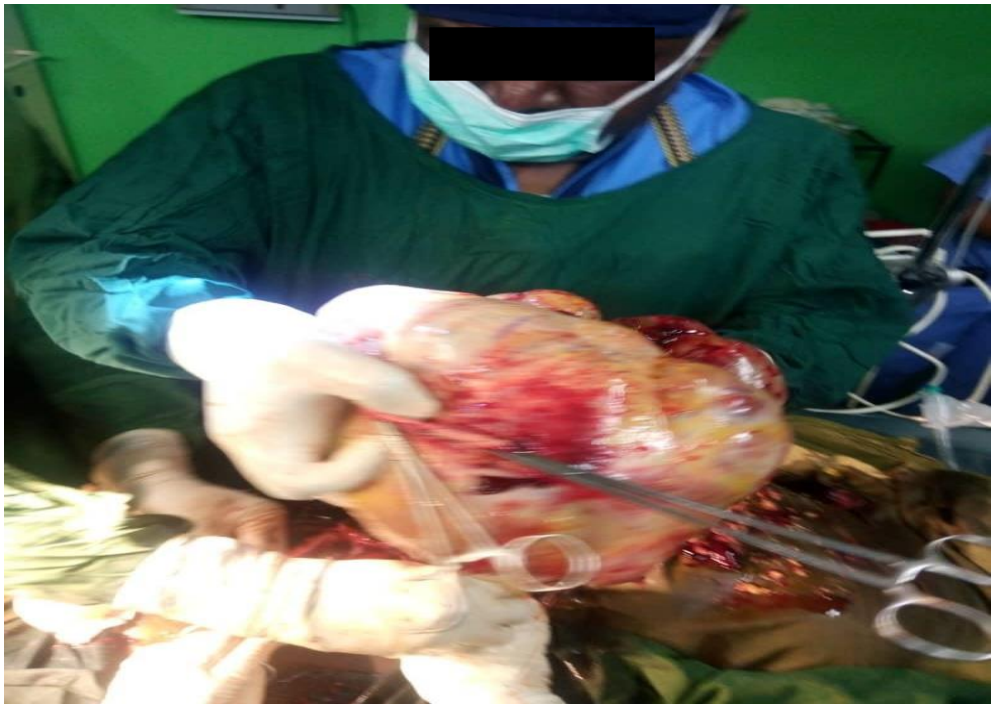

Figure 3: Tumour after removal

DISCUSSION

Ovarian tumours affect women of all age groups. In Jos University Teaching Hospital, the commonest histological variant of ovarian cancers was reported to be epithelial cancers which accounted for 55% of cases with the age at presentation of between 20-70 years.[5]

A 10 year review of ovarian tumours in Zaria, showed an age range of 8-80 years with granulosa cells such as in the reported case being the second commonest cell tumour accounting for an incidence 23.1% while serous cyst adenocarcinomas were commonest accounting for 41% of cases. [6]

In Nigeria ovarian cancers are the second commonest gynaecological cancers after cervical carcinonoma.[7,8]

The anatomical inaccessibility of the ovary coupled with the absence of a defined premalignant stage has made it difficult to develop a reliable screening technique for early detection of ovarian tumours. Most cases of ovarian tumours therefore present with advanced disease .More than 75% of cases present in advanced stages.[9,10] Generally, nulliparity, age, family history or infertility with the use of ovulation induction agents are risk factors for the development of ovarian malignancies.[11,12]

Juvenile ovarian tumours are ovarian sex cord stromal tumours which have a mean age of occurrence of 13years but 80% occur before the age of 20 and 97% before the age of 30 and rarely secrete androgens.[13] Virilization was evident in the presented patient with the deepening of her voice and clitoral hypertrophy even though there was no assay for androgens, the signs of virilization reduced 3 weeks after salpingo-oophorectomy.

Favorable prognosis is reported especially when the tumour is diagnosed in stage I such as was the case with this patient and conservation of the contralateral ovary and the uterus is sufficient treatment if they appear normal. Presentations at FIGO Stage III and above are however likely to recur and require adjuvant chemotherapy with carboplatin and etoposide.[14]

The primary treatment of JGCTs is surgical but those with advanced or recurrent disease benefit from chemotherapy and/or radiotherapy. The risk of recurrence is addressed post operatively by frequent pelvic examinations and use of tumour markers that will aid detecting recurrent disease early.[15] Optimal management protocols of these tumours have been difficult to ascertain by randomized control trials but a majority of patients with stage I disease have 10 year survival rates of 94.4%.[16]

Complete Surgical staging for this disease requires a thorough examination of pelvic and abdominal organs as was done in this patient and removal of all visible tumour. In younger patients such as the presented case who desire future fertility, unilateral salpingo-oophorectomy is sufficient. A review of 63 of such cases by Zanagnolo showed no recurrence with such conservative management with early tumours with 5 out of 11 patients becoming pregnant.[17]

CONCLUSION

A case of an 18 year old with Juvenile Granulosa Cell Tumour is presented. The clinical and surgical management was discussed and literature reviewed.

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