A Rare Case of Bilateral Malignant Sertoli Leydig Cell Tumour in Pregnancy

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Abstract
A malignant bilateral sertoli leydig cell tumour in pregnancy is a rare entity. It presents a challenging situation to the obstetrician and requires a team approach in consultation with the oncologist. 22 year old female presented to us at 5 months of pregnancy with ?fibroid of uterus. She was managed till term in ward and elective LSCS was done, when intra operatively she was found to have bilateral ovarian tumour. On histopathology it was diagnosed to be a rare case of poorly differentiated malignant sertoli leydig cell tumour which was hormonally inactive. Patient was then operated for total abdominal hysterectomy with bilateral salpingectomy with right ovariectomy with omentectomy with lymph node dissection. She was then referred to Tata hospital for chemotherapy. In present case the management has resulted in a favourable outcome in the form of a healthy mother.

Introduction
Sex cord tumours constitute about 5-7% of all ovarian malignancies, while sertoli leydig cell tumours constitute 0.2 to 0.5% of all ovarian cancers.2,3 As these tumours are rare, there is lack of proper staging, non uniform treatment and inadequate follow-up, thereby limiting our understanding of their natural history.4,5 Majority of these tumours occur in the reproductive age group (85%).2,3 50% of these cases present as solid unilateral adnexal mass with progressive features of defeminisation and virilisation,2 following a sequential event as oligomenorrhea, amenorrhea, followed by hirsutism, acne, virilization with clitoromegaly, loss of breast mass, and temporal balding.2 Few of these tumours could be hormonally inactive in nearly 10% of cases as was seen in this case.5

Case Report
A 22 year old muslim primi patient hailing from Ajamgarh was referred from private at 21 weeks of gestation in view of fibroid with pregnancy. She had no complaints at the time of admission. She gave no significant past history or history of any malignancy in her or any family member .

The patient was admitted in the antenatal ward and thoroughly investigated. On admission her vitals were stable, pulse rate was 82/mins, her weight was 48 kgs. Rest of the systemic examination was normal. On abdominal examination uterus was 22 weeks size. On per speculum examination the cervix was high up and pushed anteriorly. On per vaginal examination a firm swelling was felt in the posterior fornix occupying the pouch of Douglas and was about 10 x 10 cm in size. The clinical impression was of a subserosal uterine myoma arising from lower uterine segment or an ovarian tumour. The tumour markers (AFP, LDH, beta hCG, Ca 125 and CEA) were sent to rule out ovarian malignancy and were within normal limits.

The patient was reviewed by senior radiologist and on ultrasound was diagnosed to have a subserosal fibroid arising from the posterior lower uterine segment pushing the cervix anteriorly and an MRI was advised for confirmation. The MRI suggested a differential diagnosis of subserosal pedunculated fibroid or a complicated ovarian mass.

The doppler indices for the tumour also suggested a benign tumour and since the patient was asymptomatic a decision of conservative management...
of the case was taken in consultation with the senior gynaecologists. The patient was admitted in the antenatal ward throughout pregnancy for close monitoring, especially for the growth of the tumour. The mass increased in size from 12.5 x 11.4 x 11.1 cms to 18 x 18 cms as seen on serial ultrasound examination. Patient remained asymptomatic throughout the pregnancy. At term, the mass occupied the entire pelvis and pushed the cervix anteriorly jamming the whole birth canal. A decision to perform an elective LSCS was taken.

Intraoperatively a large bosselated mass 20 x 15 x 5 cms was noted in pouch of Douglas not separable from left ovary. The mass showed areas of haemorrhage and necrosis. Left fallopian tube was stretched over the tumour. Right ovary showed evidence of multiple hard masses largest of which was 4 x 2 x 1.5 cms. Since the patient was in the reproductive age group and the relatives were not willing for a radical surgery, left sided oophorectomy along with right-sided excision of the tumour was performed. Left fallopian tube was separated from mass and conserved. The largest of the mass of the right ovary enucleated. The specimen was sent for histopathology in toto.

The patient delivered a healthy female child of 2.4 kilos with no features of hyperandrogenism. The postoperative period was uneventful. Patient was discharged on day 5 of LSCS due to persistent request by her relatives (Fig. 1).

On histopathology at the TATA memorial hospital the tumour was diagnosed as bilateral, malignant poorly differentiated sertoli leydig cell tumour. It was termed as an unusual and rare case.

After 3 months of confirmation of report patient followed up. CT scan abdomen and pelvis this time showed enlarged right ovary 5.3 x 2.5 cms with enhancing stroma and cyst within suggesting recurrence of the previous tumour.

A detailed consultation regarding patient’s diagnosis and further management was done with oncologist. Patient was then operated for total abdominal hysterectomy with bilateral salpingectomy with right ovariectomy with omentectomy with lymph node dissection. The specimen was sent for histopathology to TATA hospital.

Histopathology report showed malignant sertoli-leydig cell tumours-poorly differentiated type in right ovary. Micro metastasis was seen in common iliac nodes. The post operative course in ward was uneventful.

She was sent to TATA hospital for further management. In Tata hospital she has been started on etoposide, cisplatin, doxorubicin and cyclophosphamide. She has been regularly following up there and now one and half years of treatment there is no recurrence on USG or CT scan.

**Discussion**

The prognostic factors include stage, histological grade (differentiation), tumour rupture and the presence of heterologous mesenchymatous constituents. The number of mitoses is a potential prognostic factor but seems to be associated with the histological grade of the tumour. In the present case the tumour was mitotically active with more than 10 /10 HPF.

Low grade tumours are very rarely bilateral (< 1%). Hence usual treatment of choice is unilateral salpingo-oophorectomy in well differentiated type with evaluation of contralateral ovary. If patients’ childbearing has been completed, total hysterectomy and bilateral salpingo-oophorectomy is performed. The tumour is not much sensitive to chemo therapy. Several drugs regimens have been tried. Alkylating drugs, adriamycin, CAP (cisplatin, adriamycin and cyclophosphamide), PVB (cisplatin, vinblastin and bleomycin). In majority of cases, the effect could not be evaluated. There is no data...
available on the effect of radiotherapy on these tumours.8

The overall 5-year survival rate is 70-90% in well differentiated type. Most fatality is seen in poorly differentiated type with 10 year survival rate of 44%.4

References

QUALITY OF LIFE WITH DEFIBRILLATOR THERAPY OR AMIODARONE IN HEART FAILURE

The use of implantable cardioverter-defibrillators (ICDs) prolongs survival for some patients with heart failure. However, it has been suggested that ICD implantation may adversely affect the quality of life. In this trial, the evaluation of several quality-of-life measures disclosed no long-term effect, although a temporary adverse effect was evident within 1 month after an ICD shock.


CIRCUMCISION TO PREVENT HSV-2 AND HPV INFECTIONS AND SYPHILIS

In two studies in Uganda involving 3393 adolescent boys and men who were seronegative for HIV and for herpes simplex virus type 2 (HSV-2), circumcision reduced the acquisition of HSV-2 and the prevalence of high-risk human papillomavirus (HPV) infection but not the acquisition of syphilis.