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A rare case of sertoli leydig cell tumour with successful pregnancy following oophorectomy

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Abstract : Ovarian SLCT are rare sex cord stromal tumours accounting for 0.1 percent of ovarian tumours. Characteristic feature of this tumour is virilisation due to testosterone secretion by Leydig cells.50 percent of patients have no endocrine manifestations. Most of the tumours are benign and unilateral with favourable prognosis following conservative surgery. One such is this case report of intermediate type SLCT in a woman who presented with virilising features and secondary amenorrhoea and underwent unilateral salphingo-oophorectomy. Post surgery she resumed regular cycles and had spontaneous conception 8 months later with successful pregnancy and good neonatal outcome.

Keyword : Sertoli leydig cell tumour, Ovary, Frozen section

INTRODUCTION:-

Sertoli Leydig cell tumours are rare sex cord stromal tumours-less than 0.1%. 75% are seen in women younger than 40yrs of age. Mean age of occurrence is around 25 yrs.DICER1 gene germ line mutation is found to be associated with SLCT. Family history of pleuropulmonary blastoma, Wilm's tumour, cervical rhabdomyosarcoma are seen in inherited cases. These tumours have testicular cells in various stages of development.30 to 50% of them are virilising.97% is stage 1 disease confined to one ovary. Just 2% spread beyond the ovary.5% have risk of recurrence or metastasis2.

CASE REPORT:-

A 22 yr old woman presented to gynaecology OPD with complaints of amenorrhoea for past 3 yrs, associated with abnormal distribution of hair growth pattern over face and chest. She had characteristic hoarse masculine voice at the time of presentation, which she noticed to develop progressively over 2 years. She was third born child to non consanguineous parents. She attained menarche at 14 yrs of age and had regular 5/30 days cycles. She had been married for 3 yrs and had normal sexual life with nil conceptions. She had no significant medical/surgical co-morbidities. Her family history was unremarkable except for deaf mutism in one of her siblings.

ON EXAMINATION:

She was thin built, tall statured with loss of feminine body contour. Her BMI was 22. She had male pattern of baldness and features of hirsuitism with Ferriman Gallway score of 13. Examination of breast showed Tanners stage 2 development. Thyroid and spine were normal. Vitals were stable. Systemic examination was unremarkable. Examination of external genitalia showed clitoromegaly[Figure 1]and Tanners stage 5 development of pubic hair. Per speculum examination showed healthy cervix. Bimanual pelvic examination revealed normal size uterus with minimal right fornicial fullness. Urine gravid index was negative for pregnancy and she was hospitalized on June 11, 2013 for further evaluation and management.

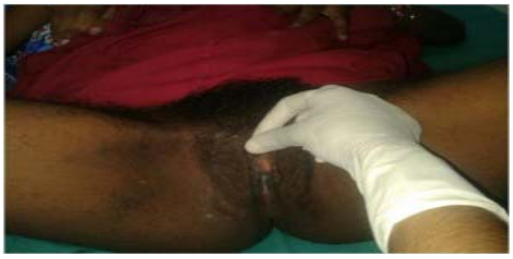

Figure 1: Clitoromegaly

BIOCHEMICAL ANALYSIS: Baseline investigations were within normal limits.

HORMONAL ASSAY:

S.Testosterone-516ng/dl

S.Free Testosterone – 2.8 pg/ml
DHEA – 2.5 ng/ml
17 OH Progesterone – 0.2 ng/ml
FSH – 6.32 mIU/ml
LH – 7.29 mIU/ml
CA 125 – 8.35 U/ml
S.Prolactin – 20.5 ng/ml

IMAGING:

TVS:- Uterus - 6.9*3.2*2.5 cm
ET – 0.3 mm

Right Ovary – 4.5*3.5*2.4 cm
Left Ovary – 3.2*1.5*1.1 cm
CT Abdomen and Pelvis:- **Oval shaped contrast enhancing mass lesion of size 4.5*3 cms in right adnexa**.[Figure 2]

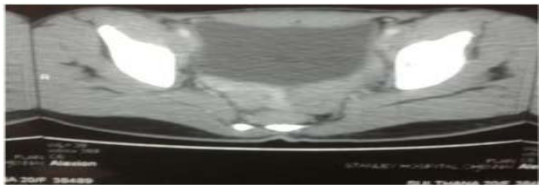

Figure 2: CT pelvis

GENETIC SCREENING:

Karyotyping showed **46XX**.

In view of hyperandrogenism, with elevated testosterone levels and right adnexal mass, She was posted for exploratory laparotomy under low risk anaesthetic fitness on 11 July 2013 with a suspicion of androgen secreating ovarian tumour.

LAPAROTOMY:

Intraoperative findings:-

Uterus – smaller than normal Right Ovary – 5*5 cm adhered to right fallopian tube. Left tube and Ovary appeared normal. No gross evidence of malignancy. Operating team decided for frozen section with wedge biopsy taken from both ovaries. Frozen section showed immature tubules interspersed in ovarian stroma in the right ovary [Figure 3]. Left ovarian wedge biopsy section was absolutely normal. No evidence of malignancy in both ovaries.

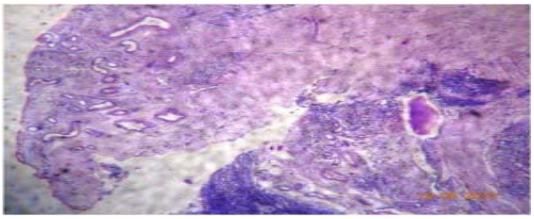

Figure 3: Frozen Section of right ovary showing primitive tubules interspersed in ovarian stroma

Intraperitoneal organs were inspected. Peritoneal wash done and proceeded with right salphingooophorectomy. Lymph node dissection and peritoneal biopsy were not performed. Resected specimen was sent for histopathology and tumour was surgically staged as IA with no preoperative spillage of tumour cells.

HISTOPATHOLOGY:

Gross specimen of right ovary appeared partially cystic with more of solid components. Surface was smooth. Cut surface showed variegated appearance with tan to brown areas of haemorrhage and yellowish areas. Microscopically, Ovarian parenchyma with neoplasm composed of lobules/hollow tubules/nests and cords were noted. Round cells with, moderate eosinophillic cytoplasm and hyperchromatic nuclei – **Sertoli cells,** interspersed with polygonal cells with abundant eosinophillic cytoplasm and round nuclei – **Leydig cells** were noted. No evidence of heterologous elements [Figure 4].

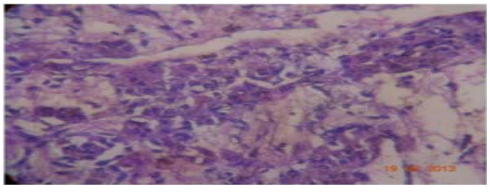

Figure 4: 40 X magnification showing Sertoli cells and leydig cells

HPE report: Sertoli leydig cell tumour of intermediate differentiation of right ovary.

On HPE, there was still a suspicion of ovatestis to be genetically evaluated. Hence **SRY gene tracing** was done by molecular genetic analysis. The test was 100% negative for SRY gene, ruling out the possibility of ovatestis and thus the diagnosis of Sertoli leydig cell tumour was explicit.

FOLLOW UP:

Patient was closely monitored for local recurrence or distant metastasis post surgery with repeat testosterone levels and ultrasound examination. Testosterone reached normal reference range within a two weeks time. She resumed her regular cycles within 20 days of tumour excision and conceived spontaneously 8 months later. She had regular AN visits, with all 3 trimesters uneventful. A week prior to her EDD, she had spontaneous onset of labour pains and delivered an alive , term , female baby of 3 kg via labour natural on 3 December 2014

DISCUSSION:-

SLCT account for 0.1% of sex cord stromal tumours of ovary. Interesting fact is that Sertoli cells and not leydig cells form the neoplastic component of these tumours. Prognosis is usually good and correlates with stage and degree of differentiation. This particular tumour appeared to be of intermediate differentiation Meyer's type 2 which accounts for 54% of Sertoli leydig cells tumours3. In a review of 207 cases by Young and Seulhy in 1985,all well differentiated tumours were benign,15% of intermediate differentiation,59% with poor differentiation and 19% with heterologous elements were malignant1.In an another study of 64 patients who had intermediate or poorly differentiated SLCT, a survival rate of 92% was noted at both 5 and 10 yrs. Most of these tumours are unilateral and diagnosed in stage 1, so conservative surgery in young patient is an appropriate treatment4. Adjuvant chemotherapy is considered for patients who have poor prognostic factors5. The patient in this report was not given adjuvant chemotherapy as the tumour was of intermediate differentiation with no heterologous elements or any evidence of metastasis of any histological type.

CONCLUSION:-

SLCT is a rare sex cord tumour that usually occurs unilaterally. SLCT should always be considered in a young female patient who has symptoms of virilization and an ovarian mass on examination or investigation. Management issues mostly revolve around the histology of the tumour. Poorly differentiated tumours require aggressive management because the chances of them being malignant are high. Intermediate differentiated tumours need an individual approach. The patient should be involved in all decision making.

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