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A CASE OF STEROID CELL TUMOR OF OVARY- CASE REPORT AND LITERATURE REVIEW

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Abstract :
OBJECTIVE - Ovarian Steroidal Cell Tumor (NOS) not otherwise specified are rare sex cord stromal tumors of ovary with malignant potential. These tumors arise in ovary and produce signs of virilization. Here we report a case of peri-menopausal female with obstructed para-umbilical hernia with signs of virilization and yellowish orange tumor was found in left ovary. METHOD - Case report and review of medical literature . CASE REPORT - A 43 year old peri-menopausal women was admitted with abdominal distension and signs of intestinal obstruction due to irreducible paraumbilical hernia. Patient was evaluated and taken up for emergency laparotomy for intestinal obstruction. Intraoperatively a segment of transverse colon with greater omentum was found inside the para-umbilical hernia sac and 1.5 litre of serous ascitic fluid was aspirated. Then thorough laparotomy was done and rest of bowel was found to be normal. Ovaries were examined as patient had signs of virilization and left ovary was enlarged 6cmx5cm in size and yellowish orange in colour with irregular surface, right ovary appeared to be normal. Bilateral oophorectomy was done. Virilization features decreased postoperatively. CONCLUSION - Ovarian steroidal cell tumors are rare and very few cases has been reported worldwide. It is a type of sex cord stromal tumor. It can be benign as well as malignant.

Keyword :
Ovarian steroid cell tumor, sexcord stromal tumor

INTRODUCTION:
Ovarian Steroidal Cell Tumor, (NOS) not otherwise specified are rare sex cord stromal tumors of ovary with malignant potential1,2. So far only a few cases were reported. Here we report a case of Ovarian Steroidal Cell Tumor. Patient presented to emergency ward with clinical signs of intestinal obstruction and obstructed para-umbilical hernia. Intra-operatively yellowish orange tumor was found in left ovary. Bilateral oophorectomy was done

CASE REPORT:

HISTORY
43 year old peri-menopausal women was admitted to emergency ward with complaints of abdominal pain for two days, vomiting and constipation for one day, patient had swelling in umbilical region for 1year which started as a small bulge and gradually progressed to present size, initially swelling was spontaneously reducible but for past two days swelling was irreducible. Patient is not a known diabetic, hypertensive or asthmatic, her last menstrual period was 6 months back.

EXAMINATION
On examination patient is well built and nourished, no pallor, not icteric, no generalised lymphadenopathy, temporal balding, male pattern of facial hairs present . Abdomen was distended with paraumbilical hernia and contents not reducible, tender, skin thinned out, free fluid abdomen was present, no organomegaly or mass palpable, bowel sounds were absent, per vaginal examination-normal, per rectal examination-empty. other system clinically normal.

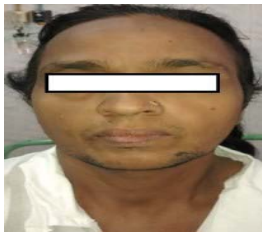

Fig 1 shows male facial hair pattern

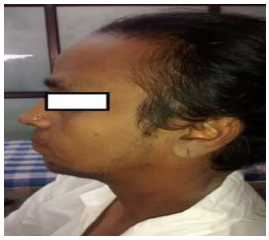

Fig 2 shows temporal balding

INVESTIGATION;
Complete hemogram, renal function test and liver function test were found to be within normal levels. Chest X-ray was normal X-ray Abdomen showed distended small and large bowel loops. With airfluid level in small bowel. USG ABDOMEN showed distended bowel loops with free fluid abdomen and pelvis not visualised due to empty bladder and foleys catheter in situ.

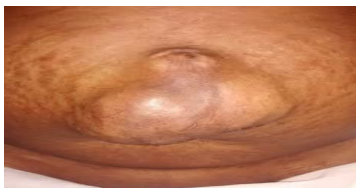

Fig 3 shows irreducible paraumbilical hernia

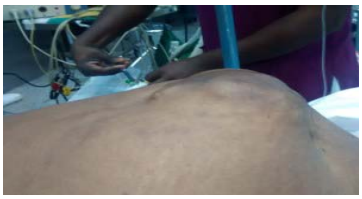

Fig 4 shows irreducible paraumbilical hernia
TREATMENT
Pre-operative diagnosis was obstructed para-umbilical hernia and patient taken up for emergency laparotomy after adequate preparation and intra-operative findings was thinned umbilical cicatrix and 2 litre of serous ascitic fluid,and the ascitic fluid was aspirated and sent for biochemical and cytological analysis, omentum and transverse colon was found as content of the hernia sac and they were viable. Thorough laparotomy done and was normal, ovaries were examined as patient had features of virilisation, left ovary was enlarged, yellowish orange in colour with irregular surface and measures 6cmx5cm in size, right ovary was normal.

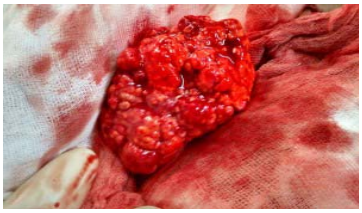

Fig 5 shows intraoperative view of ovarian tumor

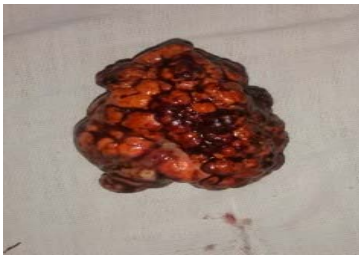

fig 6 shows ovarian tumor after surgery-yellowish orange in colour
Bilateral salphingo-oophorectomy and omentectomy was done. Mesh repair of the paraumbilical hernia using 10x10 cm prolene mesh was done. Post-operative period was uneventful . Oral feeds started on second post operative day. Patient virilisation symptoms started decreasing postoperatively. Discharged on 10th post operative day and was followed up. Ascitic fluid was negative for malignancy and histopathological examination of left ovary was steroid cell tumor of ovary (NOS) not otherwise specified.

DISCUSSION:
Steroid cell tumor of ovary ,(nos) not otherwise specified constitutes <0.1% of all ovarian tumors1. Cell of origin is not known. It is a type of sex cord stromal tumor with malignant potential3. It produces several steroids particularly testosterone. Histurism, temporal balding, amenorrhoea are seen. 1/3 of adult tumor is malignant1. Patient can present with vague abdominal pain, bloating, distension, but ascites is rare. Extensive database search was done and only 76 cases were reported since 197910. 3subtypes of steroid cell tumors of ovary11 are

- Leydig cell
- Stromal leukoma
- Not otherwise specified(rare type)

Most important predictors of malignancy are >2mitosis/10HPF, nuclear atypia, necrosis, haemorrhage, tumor size >7cm.5 In virilised patients, serum testosterone >2ng,normal DHEAS (dehydro epiandrosterone), no evidence of 21alpha hydroxylase deficiency strongly indicate ovarian virilising tumor/ovarian hyperthecosis5. IHC markers are positive with inhibin, fat stains in (75%) Vimentin in (75%) AE1/AE3,EAM(8%) S100(7%).6 IHC markers are negative with chromoganinA,CD15,myoglobin,AFP,CEA,HMB-45.6 Histopathological examination - shows usually diffusely arranged cells, also in nests, clusters, cords, columns with scanty stroma and may be filamentous, edematous or myxoid. Distinct cell border, central nuclei, prominent nucleoli. No reinke crystals (pathognomonic of steroid cell tumor, not other wise specified)4. No significant nuclear atypia/mitotic activity.

TREATMENT:
Surgery is the main modality of treatment, in adults - staging laparotomy is done, in young patientssalphingo- oopherectomy is done and follow up with serum markers2. Recently GnRH agonists has been found to be useful3.

SUMMARY:
Steroid cell tumor of ovary is a rare type of sex cord stromal tumor with malignant potential and this condition should be suspected in patients with recent history of virilisation and further evaluation and treatment should be done based on radiological, bio-chemical and pathological investigations. Our patient presented with signs of acute intestinal obstruction and ovarian tumor was found as an incidental finding as ovaries were examined since patient had signs of virilisation. Hence patient was taken up for emergency surgery, hormonal assay could not be done .Bilateral salphingo oophorectomy and omentectomy was done. Ascitic fluid was aspirated and sent for pathological and biochemical analysis. Ascitic fluid analysis was negative for malignancy. Post operatively patient had decrease in virilisation features and histopathology showed Steroid Cell Tumor of Ovary (NOS) not other wise specified. Steroid cell tumor of ovary has high malignant potential and pathological examination should be focused on finding the features suggestive of malignancy. Biochemical markers and immunohistochemical markers are essential for diagnosis. Treatment has to be specified based on patient age and histopathological report. Patient should be under strict followup.

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