HUGE TWISTED AND RUPTURED GRANULOSA CELL TUMOUR IN A PERIMENOPAUSAL FEMALE WITH HAEMORRHAGIC SHOCK - A CASE REPORT

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PRESENTATION OF CASE

Granulosa Cell Tumours (GCTs) though accounting for approximately 70% of malignant sex cord stromal tumours are rare, they comprise of only 2 to 5% of all ovarian neoplasms.[1-3] These tumours arise from granulosa cells that are hormonally active stromal elements in close association with ovarian oocytes, which are responsible for the production of estradiol.[1] Granulosa cell tumour is a vascular tumour that may occasionally rupture and result in abdominal pain, hemoperitoneum and hypotension mimicking an ectopic pregnancy in younger patients. Tumour rupture is often attributed to haemorrhagic cyst in up to 10 to 15% of the cases.[2,4]

I report an interesting case of huge adult granulosa cell tumour in a perimenopausal women who presented with acute abdomen, hemoperitoneum and haemorrhagic shock.

CASE REPORT

A 52-year-old, P3 L3, all FTNVD, LCB and tubectomy, 25 years back presented in emergency with complaints of severe pain abdomen, fainting attacks and vomiting since 1 hour. On detailed history, she gives history of irregular menses since last 6 months with amenorrhoea of 2-2½ months followed by bleeding and spotting off and on in between.

On examination, the lady was found to be in shock with vital signs of pulse rate 140 beats per minute, blood pressure systolic 80 mmHg, respiratory rate 32 breaths per minute and cold and clammy peripheries. Although, she was looking very pale, her consciousness was clear. Per abdominal examination revealed gross distention and tenderness of whole abdomen. An ill-defined mass of approximately 28 weeks size uterus was palpable in the midline. Emergency ultrasonography and CECT whole abdomen was done along with simultaneous resuscitation in ICU. Ultrasound and CECT findings revealed a large solid cystic mass arising from the pelvic cavity of about 17.5 x 16.2 x 10 cm (1425 cc volume) abutting the uterus. Areas of hyperdensities were seen in the non-contrast images? Bleed. A large heterogeneously hyperdense area was noted in superior and lateral aspect with superior extension into supraumbilical region. Both ovaries were not seen separately from mass lesion. The findings were suggestive of right-sided huge ovarian mass with moderate ascites and hemoperitoneum?.

After stabilising the patient, decision for emergency laparotomy was taken. On laparotomy, patient had hemoperitoneum of approximately 3 litres, which was drained. Patient was replaced with 4 units of PRBC, 4 units of FFP and 4 units of platelets, preoperatively. There was a huge ruptured and twisted right ovarian mass of size 18 cm x 17 cm, which was actively bleeding. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done. Imprint was sent, which showed haemorrhagic debris. Histopathology and immunohistochemistry showed granulosa cell tumour of the right ovary. Left ovary and both the tubes were unremarkable. Uterus showed adenomyosis and cervix showed chronic cervicitis. Omental biopsy and peritoneal fluid were negative for neoplastic infiltration. She was diagnosed with uncontrolled sugars during investigations and was managed adequately. She had an uneventful postoperative period and was discharged home on 6th postoperative day with a plan for oncology consultation and chemotherapy with bleomycin, etoposide and Cisplatin for 3 to 6 cycles.

DIFFERENTIAL DIAGNOSIS

Patient of perimenopausal age with gross abdominal distension, pain and shock, the first possibility of ruptured ectopy is kept in mind. However, ruptured ovarian tumour is another strong possibility, which was supported by clinical examination of approximately 26 weeks size mass in the lower abdomen in the present case.

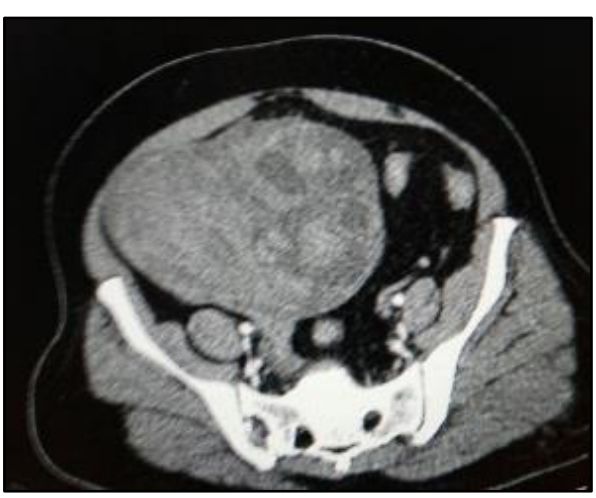

Figure 1. Huge Mass Arising from Pelvis

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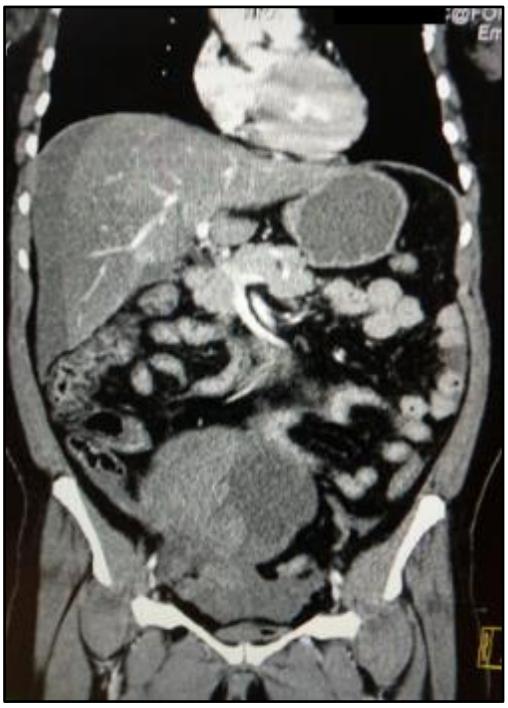

Figure 2. Under Edge of Mass Showing Breach- Indicating Rupture

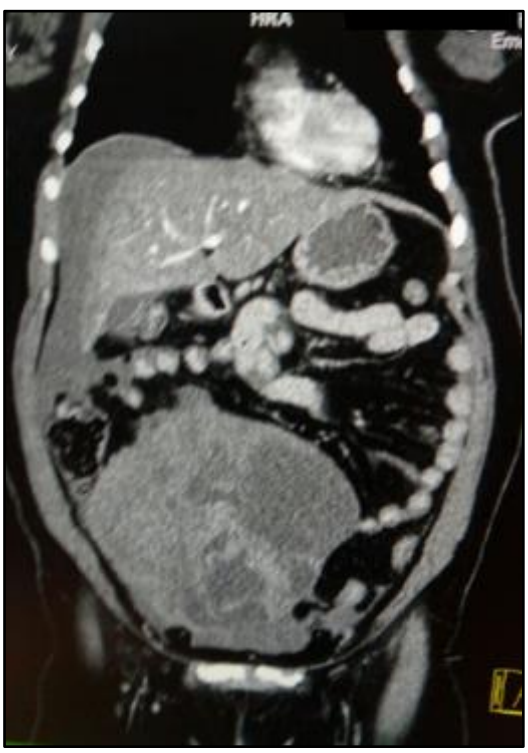

Figure 3. Gross Hemoperitoneum

CLINICAL DIAGNOSIS

On clinical history and thorough examination, the diagnosis of ruptured ovarian tumour was made, which was supported by USG and CECT.

PATHOLOGICAL DISCUSSION

Granulosa cell tumour were first described in 1855 by Rokitansky.[5] GCTs are derived from granulosa cells and are rare neoplasms, which constitute less than 5% of the ovarian tumours and more than 70% of sex cord stromal tumours.[1] There are two distinct histological types- Adult GCT (95%) and juvenile GCT (5%), which display different clinical and histopathological features.[6] They are bilateral in only 2% of cases.

No specific risk factors are identified for granulosa cell tumours. Mutation of FOXL2 gene maybe pathognomonic for Adult Granulosa Cell Tumour (AGCT).[7] Well-differentiated granulosa cell tumours have a microfollicular, macrofollicular, trabecular, insular, solid tubular and hollow tubular pattern characteristics. Call-Exner bodies are seen in 30% of cases.[8] Immunohistochemistry help clinch the diagnosis in difficult cases. Granulosa cell tumour is alpha inhibin and calretinin positive.[9] They can occur at any age and vary in size. Symptoms related to hyperestrogenism occur like irregular or heavy menstrual bleeding, intermenstrual bleeding or amenorrhoea. Endometrial cancer occurs in association with granulosa cell tumour in at least 5% of cases and 25 to 50% are associated with endometrial hyperplasia.

DISCUSSION OF MANAGEMENT

Surgical staging remains the initial management of a suspected case of granulosa cell tumour. There is no proven role of nodal dissection. The 5 to 10 year disease-specific survival was 97% and 94%.[10] Various factors shown to have prognostic significance include age, tumour size, rupture of tumour, mitotic activity, nuclear atypia, aneuploidy (in 5%-20% GCT), P-53 overexpression and stage of disease.[7-9,11-13] There is no evidence that adjuvant chemotherapy should be given routinely.[14] Women with macroscopic or microscopic residual disease (i.e. stage II-IV) Cisplatin-based chemotherapy is utilised. Most authors recommend the use of bleomycin, etoposide, and platinum (BEP) for 3 to 6 cycles.[15] Late recurrence is a hallmark of ovarian granulosa cell tumour, hence indefinite followup of patients with ovarian granulosa cell tumour has been emphasised.[16,17] Most common tumour markers useful in early detection of recurrence are inhibin B and Mullerian inhibiting substance/AMH.[18-20]

FINAL DIAGNOSIS

With history, clinical evaluation, radiological evidence and finally laparotomy and histopathology, final diagnosis of ruptured adult granulosa cell tumour of ovary was made.

Granulosa cell tumours are called multifaceted tumours. While typical cases do exist, characteristic of granulosa cell tumours in clinical practice do not always fit within the confines of these parameters. Hence, suspicion of granulosa cell tumour and gynaecological emergency should be kept in mind. In perimenopausal and postmenopausal females, prompt management and adequate resuscitation provides good results. Proper counselling and emphasising the importance of lifetime followup is imperative.

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