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A RARE CASE OF RUPTURED ADULT GRANULOSA CELL TUMOUR OF OVARY

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ABSTRACT A 37-yearold, perimenopausal woman was brought to emergency department with severe abdominal pain. Physical examination revealed acute abdomen findings with pelvic mass on the right adnexal region. Immediate exploratory laparotomy was performed. During laparotomy 1000 cc of bloodstained fluid, ruptured and actively bleeding large mass arising from right ovary was observed. Right salpingo-oopherectomy was performed in emergency conditions, and pathology report revealed an adult type of granulosa cell tumor. After this result, staging surgery was performed and patient was diagnosed as granulosa cell tumor stage 1 c followed by 4 cycles of chemotherapy was given. Clinicians should be aware of granulosa cell tumors which may occur at any age and prone to rupture. Frozen section will be helpful in order to avoid incomplete surgeries especially in women presented with intra-abdominal bleeding

KEYWORDS : Granulosa cell tumor Case report Abdominal pain Hemorrhage Ovarian cancer

INTRODUCTION:

Granulosa cell tumors are the most common type of malignant ovarian sex cord stromal tumor.Incidence 2%-5% all ovarian malignancies Granulosa cell tumor are considered to be low grade malignancy. 70% granulosa cell tumor secrete estrogen or androgen. Women may present with an asymptomatic mass or symptoms related to hyperestrogenism like abnormal uterine bleeding, breast tenderness, and postmenopausal bleeding in adult form of the disease. It is reported that tumor rupture occurred in 17.6% of cases before surgery diagnosed as adult granulosa cell tumor but the pain did not present as a cause of acute abdomen. We report a 37-year-old multiparous woman who presented with acute abdomen and hemoperitoneum because of ruptured granulosa cell tumor

Case Report:

A 37-year-old woman presented to the emergency department with history of amenorrhea, irregular menstrual bleeding, history of pain abdomen on & off since 3 weeks, history of increase in severity of pain abdomen. Past medical history was significant for T2DM ON OHA. No history of allergy or blood transfusions. Surgical history was positive for appendicectomy and one Caesarean section. Family and psychosocial history were non contributory

On examination, the patient was hypotensive and dyspneic. The abdomen was soft, diffusely tender, and distended. A large, ill-defined, firm, and fixed abdominal mass was noted. Diagnostic laboratory testing exhibited a critically low hemoglobin of 7.2 g/dL, which was treated with transfusion of two units of packed red blood cells (PRBCs). Physical exam findings of abdominal mass, tenderness, and poor vitals prompted imaging to diagnose a possible surgical emergency such as gastrointestinal perforation or intra-abdominal bleeding. Non-contrast abdominal and pelvic computed tomography (CT) scan demonstrated a large pelvic mass with significant hemoperitoneum, Findings likely suggestive of of ruptured right adnexal ectopic pregnancy with clot in pelvis with hemoperitoneum. Patient taken for emergency laparotomy proceeded to right salphingo – oopherecetomy. Intra operating findings – hemo peritoneum~1000cc of blood stained fluid.(IMAGE 1,2,3)

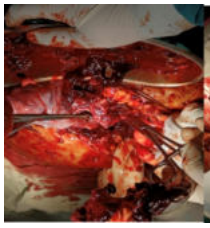

Image 1

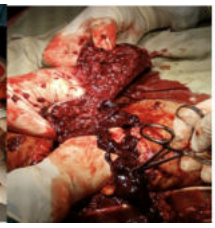

Image 2

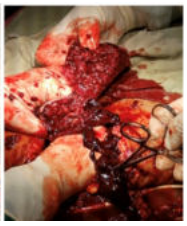

Image 3

Dense adhesions between omentum and anterior abdominal wall, bladder drawn up. Right ovary showed mass of 14 x 10 cm, cystic with evidence of ruptured, right fallopian tube walls adherent and stretched over the mass and torsion twice the time noted at the pedicle. Left ovary and fallopian tube normal. Uterus normal. Surgeon call over given, bowel walkthrough done, small and large bowel found to be normal. Peri operative 2 units packed rbc, 4 units of FFP transfused intra operatively. Postoperative period was uneventful.. Histopathology report showed Adult granulosa cell tumor of right ovary, demonstrates nuclear grooves or nuclei with a “coffee bean appearance as well as Call-Exner bodies.(IMAGE 4)

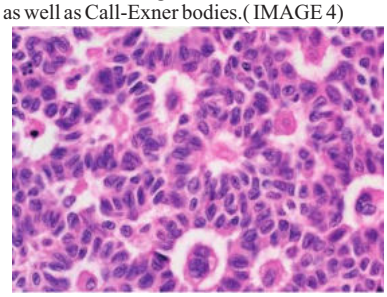

Image 4(call Exner Bodies)

Or endometrial adenocarcinoma in untreated patients. Ultrasound patient Planned for completion staging Laprotomy . Milliarypelvic nodule present. Left ovary enlarged and thickened. Entire bowel walking done, no nodules present, all solid organs normal, pelvic nodule of 6 cm with adhesions between rectum,uterus,bladder, pelvic side wall and omentum. Adhesiolysis and omentectomy done, deemed inoperable due to dense adhesions.Specimen sent for HPE. HPE showed no evidence of nuclear atypia.patient underwent completion surgery at Cancer institute (adyar- Chennai). Post operative period was uneventful followed by four cycles of chemotherapy given

DISCUSSION:

Granulosa cell tumors are sex cord-stromal cell tumors derived from granulosa cells that account for only 2–5 % of ovarian tumors . Adult GCTs present during the postmenopausal period with a median age of 50–55 years . These indolent tumors generally grow slowly and present with nonspecific symptoms, allowing them to go unnoticed for years. Symptoms are related to mass effect and excess estradiol exposure. The most common initial manifestations of ovarian GCT are abnormal vaginal bleeding, pain, and distension. Prolonged exposure of endometrium to tumor-derived estradiol may result in endometrial hyperplasia is most often used to confirm the physical exam findings and to measure the mass. CT imaging is useful for evaluation of suspected GCT, particularly in the acute setting. Definitive diagnosis is confirmed by biopsy and histopathologic analysis after surgical removal. This case describes a spontaneously ruptured ovarian GCT and acute life-threatening bleeding requiring aggressive resuscitation and prompt surgical exploration in a perimenopausal patient. The differential diagnosis for spontaneous intra-abdominal bleeding includes splenic rupture, ruptured ovarian cysts, ectopic pregnancy, ruptured abdominal aorta aneurysms and visceral artery aneurysms, and pancreatitis. Due to the lack of specificity in clinical symptoms, it is difficult to arrive at a definitive diagnosis prior to surgery. The

patient presented here was particularly difficult to evaluate due to body habitus. Laparotomy with complete tumor removal and histopathologic confirmation of the diagnosis, along with standard surgical staging, is the recommended treatment. Characteristic histology for GCT demonstrates nuclear grooves or nuclei with a "coffee bean appearance as well as Call-Exner bodies. Calretinin, a proposed marker of GCT, is used in pathohistological identification. Premenopausal women presenting in reproductive years with early-stage disease are managed with tumor resection and unilateral salpingo-oophorectomy to preserve fertility. In post-menopausal women and those who have completed childbearing, a total abdominal hysterectomy and bilateral salpingooophorectomy are recommended Despite the slow growth and silent nature of these tumors, most patients are diagnosed at an early stage. The key prognostic factor is the stage of the tumor at the initial surgery. Those with low-risk stage I GCT should be observed following surgery. Patients with GCT greater than 10 cm, tumor rupture, poorly differentiated tumor, and high mitotic index are high-risk and should consider adjuvant chemotherapy to reduce the risk of recurrence. Stage II-IV GCTs should be treated postoperatively with chemotherapy. Adjuvant radiation therapy in all stages of GCT is controversial, and its use is most often palliative for diffuse intra-abdominal and metastatic disease. There is no standard approach to recurrent GCTs; however, surgical removal may provide long-term disease control. Life-long follow-up with tumor markers, such as inhibin B, and periodic CT scans is strongly recommended regardless of stage as GCTs have greater than 40 % recurrence rate and potential for late relapse. Ten year survival rate is 84–95 % for stage I tumors, 50–65 % for stage II, and 17–33 % for stages III and IV [4,7,9]. Early recognition and surgical removal of GCT is critical to avoid unnecessary morbidity and mortality

CONCLUSION :

Granulosa cell tumors grow to be highly vascular, making them prone to hemorrhagic rupture [3]. Patient presentation of a hemorrhagic GCT is nonspecific and does not always present as hemorrhagic shock.

Associated with low grade endometrial cancer in 5% cases and Endometrial hyperplasia in 25%-50% cases Surgery alone is sufficient primary therapy. No evidence that adjuvant radiotherapy and chemotherapy will prevent recurrence and improve prognosis. Radiation and chemotherapy are reserved for treatment of recurrence or metastatic disease.

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