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Gynaecology

PELVIC MASS: DARE BUT HANDLE WITH CARE!

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ABSTRACT 75 years old housewife presented in OPD with acute abdominal pain. She had h/o abdominal pain since 1 month and abdominal distension and asymmetrical pedal edema more on left side. She had Diabetes Mellitus and past history of vaginal Hysterectomy done 25 years ago for prolapse uterus. On evaluation there was a Firm pelvic mass of 30 weeks palpable.
CT Scan showed A solid cystic mass with internal septae and wall thickening likely ovarian malignant tumor.CA 125 (21), USG RMI = 189 score. However, she also had right pulmonary artery thrombus and DVT of left lower limb.
After taking high risk consent with counselling patient was operated. Intra-Operatively ovarian mass seen to be benign ovarian tumor possibly mucinous cystadenoma. However, H.P.R. s/o Adult Granulosa Cell tumor. Anticoagulation started after 12 hours of surgery. Patient was shifted to warfarin and discharged. Dilemmas and difficulties in preoperative, intraoperative and postoperative management are discussed.

KEYWORDS : adult granulosa cell tumor, dilemmas and difficulties, pulmonary artery thrombus and DVT

INTRODUCTION

Adnexal masses represent a spectrum of conditions from gynecological and nongynecological sources. Chances of undergoing surgery for ovarian mass in their lifespan is 5 – 10%. Out of these women 13% - 21% suffer from malignancy.[1] Two percent of all ovarian cancers occur in young female (<25 years).[2] Adult granulosa cell tumors account for approximately 1% to 2% of all ovarian tumors and 95% of all granulosa cell tumors.[3] They occur more often in post-menopausal than pre-menopausal women, with a peak incidence between 50 and 55 years of age. Granulosa cell tumors (GCTs) of the ovaries originate from sex cord stromal cells. Based on histological appearances they are further classified as adult cell granulosa cell tumor and juvenile granulosa cell tumors (JGCTs).

CASE REPORT

A 75 Years old female came with chief complaint of abdominal pain and distention since 1 month, swelling of left lower limb since 1 week. She had Obstetric history – P4 L4 (all home deliveries), tubal ligation done 35yrs back and hysterectomy 25 years back (for prolapse uterus). The patient was known case of diabetes mellitus from 10 years and on oral hypoglycemic drugs. On Per abdomen and Per vagina examination 30 weeks solid cystic mass was felt and per speculum showed healthy vault.

Initial laboratory workup revealed anemia with a hemoglobin of 8.6 g/dL. CA 125 level was 21.8 U/mL. On Ultrasonography large complex cystic mass of multiple septae with low level echo of size 22.7x20.0x12.0 cm was seen and RMI index of 189. CT scan showed solid cystic mass reaching almost up to the gall bladder with ovaries not visualized separately and showing thick enhancing solid components with internal septae and wall thickening likely ovarian malignant neoplasm. Hydrosalpinx seen in relation to right adnexa. Right pulmonary thromboembolism seen. CT Pulmonary angiography showed pulmonary thromboembolism with mild thrombus in right pulmonary artery extending to proximal right lobe and lower lobe artery. Colour Doppler of lower limbs showed diffuse subcutaneous edema of left lower limb, Deep Venous Thrombosis noted in proximal Superficial Femoral Vein and Great Saphenous Vein with no Colour flow seen in left Common Femoral Vein, Superficial Femoral Vein & popliteal vein. 2D Echo showed a LVEF of 60%, trivial MR, sclero-degenerative affection of aortic and mitral valve

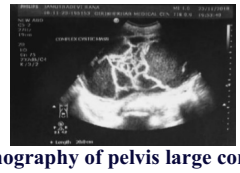


Figure 1: Ultrasonography of pelvis large complex cystic mass of multiple septae

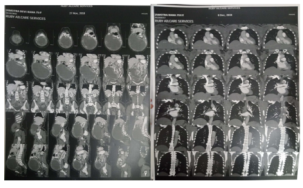
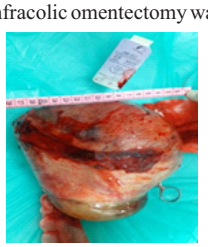


Figure 2: CT scan of abdomen and pelvis and CT Pulmonary Angiography

Medical opinion suggested 6 weeks of thrombolytic therapy and then to take up the case for surgery. However, having a large, complex cystic mass on imaging, possibly malinant and probably causing deep venous thrombosis decision to operate after high risk informed consent taken. After bowel preparation, Explorative Laparotomy was done as per standard protocols. A large solid cystic mass of approximately 20x20x10 cm size noted, freely mobile arising from left ovary reaching up to gall bladder. Mass was twisted with one and half rotation. Right and left side hydro salpinx was noted. Left ovary atrophic. Minimal ascites. Palpation of liver was normal. No gross lymph node metastasis seen. Gall bladder found to be enlarged with stones. A bilateral salpingoopherectomy with appendicectomy with infracolic omentectomy was done.



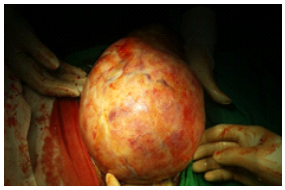


Figure 3: Intraoperative findings showing a large solid cystic ovarian mass

Postoperatively sutures were healed well. Patient was given LMWH twice a day overlapped with warfarin initially in postoperative period and warfarin in postoperative period. The leg swelling reduced after surgery. Patient was discharged after 7 days on warfarin 3 mg OD, with PT/INR maintained at 2-3. Patient was followed up after 1 month postoperatively. Wound healthy. leg swelling was completely reduced. Medicine management advised to continue tab warfarin 3 mg OD with PT/INR maintained at 2.

Histopathology report revealed Multiple section from ovarian cyst showed cellular tumor with areas of necrosis with focal Call-Exner Bodies s/o adult granulosa cell tumor. Bilateral fallopian tubes unremarkable. Appendix shows features of obliterative appendicitis.

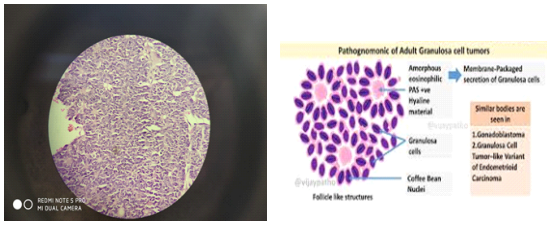


Figure 4: Histopathological slide showing call-exner bodies s/o adult granulosa cell tumor

DISCUSSION

Dilemmas and difficulties in our case,
Pre-operative - pre-operative fitness
- optimal time for surgery
Intra-operative - high risk surgery
- extent of surgery
Post-operative - complications associated with pelvic surgery and deep vein thrombosis
- decision of radiotherapy and chemotherapy

Granulosa cells are the somatic cells of the sex cords of the ovary which are closely associated with the developing oocyte. Granulosa cells differentiate from either the coelomic epithelium or mesenchymal precursors (the embryological origin is still disputed). The major functions of granulosa cells include the production of sex steroids and various peptides required for folliculogenesis and ovulation.

Granulosa cell tumors are different from the epithelial ovarian cancers on the basis of nature of presentation and clinical behavior. They occur in a younger age group, are usually detected in an early stage and often have features of hyperestrogenism. They are more readily cured by surgery alone. Generally, they have a better prognosis than epithelial ovarian tumors and follow an indolent course. They are characterized by a long natural history and 25 % may recur years after apparent clinical cure of the primary tumor.

Similar to epithelial ovarian cancers the presenting symptoms are usually nonspecific with abdominal pain (41.1 %) or distension (26.4 %).[4] These patients present with a large palpable ovarian mass. In JGCT abdominal pain was seen in 28.3 % and abdominal mass in 45.5 %.[5]

Symptoms related to hyperestrogenesim occur in all age groups. In prepubertal age group, precocious puberty with breast development increased pubic hair, vaginal bleeding (27.3 %) [5] and increased growth will be seen. In the reproductive age group, altered menstrual patterns (32.8 %)[4] like menorrhagia, intermenstrual bleeding or amenorrhea may manifest. Postmenopausal bleeding is the most common finding in the postmenopausal age group. The high estrogen levels may have contributed to the thrombosis in our case.

Around 25–50 % cases are associated with endometrial hyperplasia and endometrial cancer is seen in 5–13 % cases. Endometrial cancers, usually detected in the pathological specimens, are well differentiated early stage disease and have a good prognosis. Primary uterine cancer with ovarian metastasis or synchronous endometroid ovarian and uterine cancer has to be excluded in cases of uterine adenocarcinoma with an ovarian mass. Breast enlargement and tenderness occurs secondary to estrogen action.[6] Because of the high vascularity, tumor rupture is seen in 10 % cases, and presents with acute abdominal pain, abdominal distension and hypotension due to hemoperitoneum.[7]

Unlike epithelial tumors, GCT presents at early stage in 81 % cases (stage I-71 %, II-10 %) and at late stage in 19 % (III-11 %, IV-8 %).[8] In advanced stages, the pelvis, intra-abdominal organs and peritoneum are involved. It's unusual for patients to present with pulmonary and skeletal metastasis.

Radiologically GCT can appear either as a solid mass, a mass with hemorrhagic or fibrotic changes, a multilocular cyst or a completely cystic tumor. Unlike epithelial tumors GCT are usually confined to the ovary at diagnosis, don't have intracystic papillary projections. The incidence of peritoneal spread is low and they are rarely bilateral. Enlarged uterus with a thickened endometrium is noted due to estrogenic action. Ascites is very rarely seen.[9,10]

Differential diagnosis of patients with adnexal mass and abnormal vaginal bleeding
1. Primary uterine cancer with metastasis to the ovary.
2. Primary ovarian cancer with metastasis to the endometrium.
3. Synchronous ovarian and endometrial cancer

Surgical staging remains the initial management of a suspected case of GCT. The principles of surgery are similar to epithelial ovarian tumor with a vertical midline incision. Surgical staging includes exploration of peritoneal cavity, washings for cytology, multiple peritoneal biopsies and omentectomy

Several studies[11-13], evaluating the role of lymph node dissection have shown that nodal dissection is not a significant factor for survival and is not recommended in surgical staging of GCT. Enlarged or suspicious nodes should be removed to allow evaluation and maximal cytoreduction. In the series by Brown et al. [12] none of the 58 patients who had nodal sampling had positive nodes.

In patients where fertility is not an issue, a total abdominal hysterectomy and bilateral salpingo-oophorectomy with removal of all gross disease is the definite initial treatment. Fertility preserving surgery with unilateral salpingo-oophorectomy is feasible in young patients with stage IA GCT. The results of various studies have shown that, there is not much difference with a conservative approach when compared to the radical surgery 97 % vs. 98 % respectively. The 5- and 10-year disease specific survival was 97 % and 94 %.[7] As the incidence of bilateral disease is low (2 %) [4] a wedge biopsy of the opposite ovary is controversial and must be done with caution.

GCT have a tendency for late recurrence. Once the tumor recurs it's fatal in 80 % cases. In the study by Park et al.[14], none of the patients with early stage disease who underwent optimal debulking had tumor recurrence and of the patients with advanced stage GCT who received at least 6 cycles of BEP adjuvant chemotherapy none had tumor recurrence.

The longest reported time to recurrence is 40 years.[15] About 21 % develop recurrence and the median time to relapse was 57.6 months (2–166 months) as reported by Sun et al.[16]

Local pelvic recurrence is reported in 70 % cases, 9 % in pelvis and abdomen, 6 % retroperitoneum, 6 % pelvis and retroperitoneum and 3 % pelvis, abdomen and retroperitoneum (Abu-Rustum et al.).[17] Most recurrences are intraperitoneal suggesting the possibility of missed peritoneal disease during primary surgery especially for early stage disease. According to a study by Fotopoulou et al. [13] the tumor dissemination patterns differed significantly between primary and recurrent patients, having significantly higher rates of diffuse peritoneal involvement and extraovarian tumor involvement of the middle and upper abdomen in the recurrent cases. Not surprisingly, only about 85 % of the relapsed patients could be operated without residual lesions compared to nearly 100 % in all primary patients. Multivisceral involvement with metastasis to liver, appendix and intestines are quite common. Metastases to lung, bone, vagina, adrenal, spleen, pancreas, gall bladder, rectus muscle are rarely reported.

CONCLUSION

This rare case of postmenopausal adult GCT with its medical complications posed many challenges, but ended with good outcome. Adult granulosa cell tumours have a better prognosis than most other ovarian malignancies, stage of disease being the most important prognostic factor. Surgical clearance of the tumour is the main-stay of treatment. Emphasis must be laid on the need for long term follow-up

in view of recurrences that occur several years after the initial disease. Emergence of pelvic mass after hysterectomy poses diagnostic and therapeutic challenges to gynecologist. Decision to perform oophorectomy should be individually based taking into consideration age, family risk factors.

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