Granulosa cell tumor complicated by torsion, rupture and hemoperitoneum

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INTRODUCTION

Granulosa cell tumors account for approximately 1% to 2% of all ovarian tumors and 70% of all sex cord stromal tumor.1 It has a reported incidence of 0.5-1.5 per 100,000 women per year.2 It is mostly seen in peri-menopausal age group with a peak incidence between 50 and 55 years of age.3 Bilateral disease is observed in only 2% of patients. It is hormonally active tumor producing estrogen. Presenting manifestations are abdominal pain or swelling, menstrual irregularities or amenorrhea and postmenopausal bleeding.4 At times torsion and tumor rupture can complicate causing the patient to develop acute abdominal pain, abdominal distension and hypotension due to hemoperitoneum.5 We reported this case of 65 year old postmenopausal woman with torsion of granulosa cell tumor because of rarity of such presentation.

CASE REPORT

65 years old para 3 from a village in Nepal presented to OPD with irregular vaginal bleeding, progressive abdominal pain and abdominal distension of 11 months. She had no significant past medical or family history. On examination, she was pale. A firm to hard irregular tender mass of 14-16 weeks gravid size was felt in hypogastrium and bilateral iliac fossa with restricted mobility. CA-125 was raised by four folds. Views from USG were a unilateral 10×8 cm complex solid-cystic mass with increased vascularity with likelihood of malignancy. Whereas CT scan showed omental caking and gross ascites. Endometrial biopsy showed weakly proliferative endometrium. After blood transfusion and correction of anemia, staging laparotomy was planned. 2 l of hemorrhagic peritoneal fluid and a 20×20 cm left ovarian tumor with bluish black breached capsular surface due to rupture, twisted once on its pedicle was seen and managed by total abdominal hysterectomy and bilateral salpingo-oophorectomy (Figure 1). On cut section multiloculated, fleshy solid areas with intracystic hemorrhage and areas of necrosis were seen which on histopathology was proven to be granulosa cell tumor (Figure 2).
Figure 1: Gross appearance of the tumor showing torsion once around its pedicle.

Figure 2: Histopathologic appearance of the tumor showing diffuse sheets of tumor cells with hyperchromatic pleomorphic nuclei with inter nuclear grooving.

DISCUSSION

Granulosa cell tumor of the ovary is a rare neoplasm that originates from sex cord stromal cells characterized by prolonged natural history, tendency to late recurrences and favorable overall prognosis. It is divided into adult type and juvenile type depending on the clinicopathological findings, adult type being 95% of cases. It can manifest as an asymptomatic mass or symptoms related to hyperestrogenism like abnormal uterine bleeding, breast tenderness and postmenopausal bleeding. Granulosa cell tumor presenting as ovarian torsion is not common. Though there are few reports in juvenile type of tumor, it is very rarely seen in adult type. Tumor rupture is seen in 10% cases which manifest as abdominal pain and hemoperitoneum. Imaging findings in adult GCT vary widely and range from solid masses to tumors with varying degree of hemorrhagic or fibrotic changes, to multilocular cystic lesions to completely cystic tumors. Tumor markers used in GCT are estradiol, inhibin, follicle regulatory protein and mullerian inhibitory substance. On gross appearance, it is a pale yellowish large unilateral cyst ranging from a few mm in size to 30 cm. Two characteristic features of granulosa cell on histopathology are grooved nuclei and Call exner bodies. Surgery is the primary choice of treatment which alone provides cure in cases with disease confined to the ovaries. However, platinum-based combined chemotherapy regimen is advised in cases with high-risk factor or more advanced disease. Five year survival rates usually are 90-95% for stage I tumors compared to 25-50% for patients presenting with advanced-stage disease. Staging is the most important prognostic factor though other factors like large size (>15 cm), bilaterality and tumour rupture can impact greatly on the survival. Despite the good overall prognosis, long-term follow-up always is required in patients with granulosa cell tumors.

CONCLUSION

Granulosa cell tumor is a rare tumor of ovary. Because of limited study, most cases remain undiagnosed even by histopathology. It should always be considered in a woman who presents with postmenopausal bleeding and co-existing ovarian tumor. Early diagnosis makes for a favorable prognosis where surgery alone is sufficient primary therapy. Radiation and chemotherapy are reserved for the treatment of recurrent or metastatic disease.

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