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CASE REPORT

Juvenile granulosa cell ovarian tumor- a rare presentation

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Abstract

Juvenile granulosa cell tumor is a rare gynecologic malignancy. A fourteen year old girl was brought to the emergency with acute abdominal pain with distension of abdomen and admitted in surgical unit, clinically suspected perforation of gut. Transabdominal ultrasonography revealed a right adenexal irregular mass with a moderate amount of ascitic fluid. On laparotomy, it was suspected malignant ovarian tumor stage Ic, and then ipsilateral salphingo-ophorectomy was done considering the age of the patient. It was juvenile Granulosa cell tumor which confirmed by histo-pathologically. The epidemiology, natural history, presentation, histological and imaging appearances, prognosis and treatment of this malignancy were reviewed.

Key words: *Juvenile, granulosa cell, tumor*

Introduction

Juvenile granulosa cell tumors (JGCT) are rare sex cord stromal tumors. Granulosa cell tumors (GCT) are encompassing 1.5% of all ovarian tumor and 6% of its malignant tumors.[1] The juvenile type accounts for only about 5% of all GCT.[2] Only 5% of GCT cases are observed before puberty and more than 60% cases occurring after menopause.[2] The terms are adult and JGCT because the tumors are not defined by the age groups in which they primarily occur, rather they are distinguished by their histological appearance, that correlates to large extent with the age of the patient.[2] This patient aged 14 years with presentation of acute abdomen needed high index of suspicion to have a such tumor and correct diagnosis will be necessary for selection of proper treatment.

The case

A fourteen year old unmarried and school going young girl was presented to the emergency with acute abdominal pain with distension of abdomen for 5 days. She got admitted in surgery unit. She had been previously healthy and with history of regular menstruation. On physical examination, she had below average body build, mildly anaemic, temperature normal, no palpable lymph node. Abdomen was hugely distended, no palpable lump. Clinically, perforation of gut was suspected at first. Transabdominal ultrasound of whole abdomen was performed- there was a

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multilobulated cystic mass about 11.5 cm ×7 cm in right ovarian region with moderate amount of peritoneal fluid. All other parameters were normal, plain X-ray abdomen AP view in erect posture was normal. Provisional diagnosis was malignant ovarian tumour. So planned for exploratory laparotomy and proper counseling of the party was done. As soon as peritoneal cavity was opened about three liters of yellowish watery mucous fluid came out. Left ovary, uterus, left fallopian tube were healthy, there was a multilobulated cystic mass about 12 cm × 7 cm in the right ovary. Right fallopian tube was distorted and attached with the mass; capsule of the tumor was ruptured in one area from which fluid came out. There was no seedling in pouch of Douglas or peritoneal cavity, no enlarged lymph node, peritoneal fluid was sent for cytological examination which was negative for malignant cell. Staging was done. Then right sided salphingo-ophorectomy was done. Post operatively patient improved gradually. Histopathology confirmed the diagnosis JGCT. There was no facility for frozen section biopsy. The final diagnosis was JGCT, FIGO stage Ic. After discharge, she was referred to oncologist. Up to the time of case reporting, she was healthy and had no recurrence.

Discussion

Ovarian cancer is the third most common neoplasm of the female genital tract, after carcinoma of the cervix and endometrium.[3] Based upon their cell type of origin, primary ovarian malignancies are classified into surface epithelium, germ cell, or sex cord tumors. Approximately 70% of sex cord tumors are GCT.[4] By definition, a neoplasm must contain a granulosa cell component of at least 10% on any one slide to be diagnosed as GCT.[5] GCT produce inhibin, a peptide hormone, which is used to monitor patients for recurrence and to evaluate the success of on going therapy.[2]

JGCT was first described in 1979.[6] It has a distinctive histological appearance and characteristically occurs in children and young adults, accounting for about 85% of GCT observed before puberty.[7] Ninety seven percent of the patients are less than 30 years of age.[1] JGCT is seen at any age of infant to more ages.[8] Nona et al. described a case of seven month old infant.[8] Sivasankaran et al. described of ten months old infant.[9] Provided an etiological factor in adult types such as increasing in the level of gonadotropins has no role on juvenile type.[8] It too occurs in both ovary and testis. These tumors are often hormonally active.[4] Clinically, these patients typically present with sign of hyper-estrogenism- precocious puberty, menstrual irregularities, post menopausal bleeding. In addition, patient may complain of abdominal pain or an abdominal mass.[1] Gross appearance of these tumors- bilateral at 2%, ruptured in initial discovery in 10%, ascities in 10% of cases, size ranges 3-32 cm, appearance may be solid and cystic neoplasm, cysts containing hemorrhagic fluid, solid is of yellow-gray color with extensive necrosis or hemorrhage.[1] Fig. 1 shows some histological changes of ovarian follicles in JGCT such as presence of lutienization, cells with immature nuclei, atypia and sign of high mitotic rate. Call-Exner bodies grooved with pale, round nuclei are the classic features of the more common adult type tumor, and the lack of those features also helps to distinguish the juvenile type from adult type.[10, 11]

Imaging characteristics of adult and juvenile GCT are non-specific and these tumors can not be reliably distinguished from other ovarian neoplasm on imaging alone. They have multiple septations which are thick and irregular.[11] Metastasis, though rare at initial presentation, appears as cystic liver masses or peritoneal implants, similar to epithelial ovarian neoplasm.[11]

The staging system used for GCT is that applied to other ovarian epithelial tumors, the FIGO staging. Staging determines which patients are at risk for recurrence and those that require chemotherapy. At presentation, 90% of JGCT are FIGO stage I, confined to

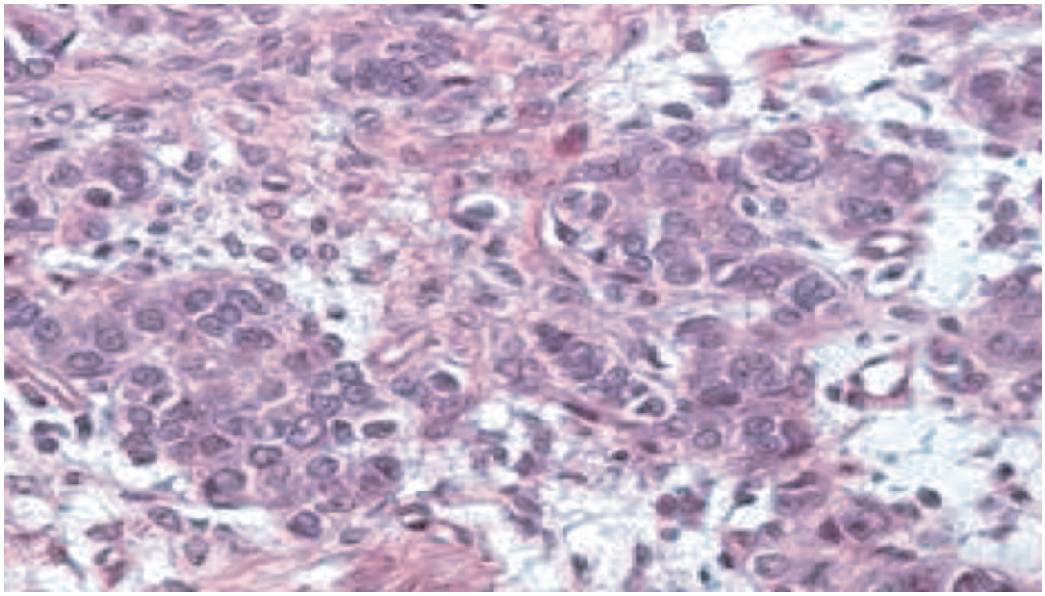

Fig. 1. Histopathological slide of juvenile granulosa cell tumor of ovary shows nuclei are immature show atypia and a high mitotic rate.

the ovary. This stage caries very favorable prognosis and a 5 year survival rate of 90-100%.[11] JGCT is less likely to occur after resection of tumor in stage I, but if recurs, is more likely rapid & aggressive with early relapses and poor outcomes.[11]

A variety of chemotherapy regimens have been used against JGCT with varying success. Multiple studies suggest that cisplatin based multi-drug regimens can be effective in advanced stages of JGCT or to treat recurrences.[12,13] Resection of stage I tumors is felt to be curative, 5 year survival for stage II tumors drops considerably to 55-75% and more so for stage III & IV tumors up to 22-50%.[14]

Unfavorable prognostic factor may include nuclear atypia, high mitotic rate, and extra capsular extension of tumor within ovary, tumor rupture and presence of residual disease after surgery.[1,12,13] Tumor markers such as inhibin can be used to assess for recurrences.

As the majority of these tumors are diagnosed at stage I, their treatment remains surgical. The role of chemotherapy and/or radiation therapy remains unclear. Given the possibility for recurrence, long term follow up and surveillance is needed for all patients.

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