Case Report

A Rare Case of Extraovarian Granulosa Cell Tumor Presenting as a Retroperitoneal Mass

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Keywords: *Granulosa Cell Tumor, Extraovarian, Retroperitoneal*

ABSTRACT

Introduction: Granulosa cell tumors (GCT) are the most common malignant sex cord—stromal tumors of the ovary. Rarely, they can occur at extraovarian site with very few cases mentioned in literature.

Case history: A 45 yr old lady presented with pain abdomen and on ultrasonography, bilateral ovarian cyst was found. Patient underwent total abdominal hysterectomy with bilateral salpingoophorectomy and during surgery, a retroperitoneal mass was discovered which was excised. On gross examination, the mass measured 7x5x5cms, was solid with areas of hemorrhage and focal areas of necrosis . Microscopy showed solid sheets of cells with round to ovoid nuclei and scanty cytoplasm. Some of the cells showed nuclear grooves. A diagnosis of extraovarian adult granulose cell tumor was madeand immunostaining with inhibin was positive.

Conclusion: Extraovarian granulosa cell tumor needs to be kept in mind in a female patient with a retroperitoneal mass. This case is presented due to its rarity and also to emphasise the fact that these tumors are often missed in the pre operative evaluation of patients and need histopathology for proper diagnosis.

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Introduction

Granulosa cell tumors (GCT) are the most common malignant sex cord—stromal tumors of the ovary. Histopathological examination of GCT distinguish two subtypes: an adult type GCT that is found typically in older women and a juvenile type GCT that is recognized primarily in children and young adults.[1]

Rarely, GCT can develop at an extraovarian site. In the English language literature, only a few numbers of cases have been presented so far. We present a case of a 45 year old female presenting with a retroperitoneal mass which was diagnosed as granulosa cell tumor on histopathology. To the best of our knowledge, this is the 4th reported case of extraovarian granulosa cell tumor from India.

Case Report

A 45 year old lady presented to the gynaecology OPD with chief complains of pain in the abdomen which started 6 months back. Ultrasonography revealed bilateral ovarian cyst. Routine blood examination was within normal limit. Surgery was performed for ovarian cyst during which a retroperitoneal mass was noted, separate from the ovaries. Patient underwent total abdominal hysterectomy with bilateral salpingoophorectomy along with removal of the retroperitoneal mass. A provisional diagnosis of retroperitoneal sarcoma was made and the specimen sent for histopathological examination. On gross examination, bilateral ovaries were cystic containing serous fluid. An irregular tan coloured mass was received measuring 7x5x5cms. Cut section was solid with areas of hemorrhage and focal areas of necrosis. [Fig.1] Microscopic examination of sections from the ovaries showed serous cysts. Endometrium showed mild endometrial hyperplasia. Sections from the mass showed solid sheets of cells with round to ovoid nuclei and scanty cytoplasm. Some of the cells showed nuclear grooves. Few mitotic figures were noted [Fig 2.3] A diagnosis of extraovarian adult granulosa cell tumor (diffuse type) was made on the basis of histomorphologic features. Immunohistochemistry showed positivity for inhibin confirming the diagnosis. The patient is currently on follow up.

Discussion

Granulosa cell tumors (GCTs) comprise 2%-5% of all ovarian tumors . [2]

Recurrence or metastasis can be seen many years after initial treatment. Primary extraovarian GCT is an extremely rare tumor. [3]

Due to the origin of sex cord from mesonephros, the ectopic gonadal stromal tissue is believed to be responsible for the histogenesis of extraovarian sex-cord stromal

Fig. 1: Showing tumor mass along with the resected specimen of uterus and ovaries.

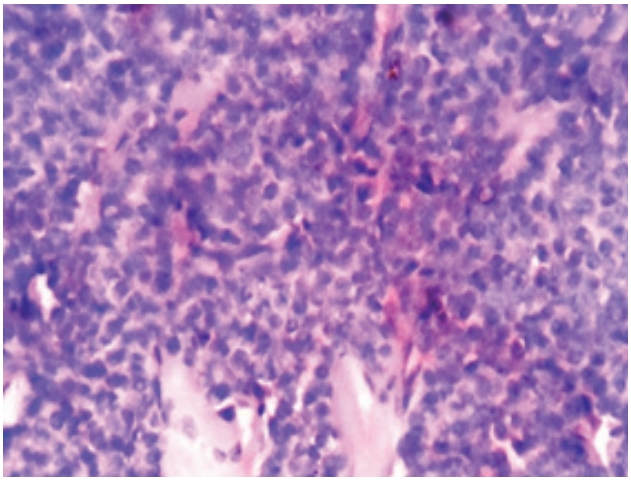

Fig. 2: Showing sheets of neoplastic granulosa cells in diffuse pattern(H&E, 40X)

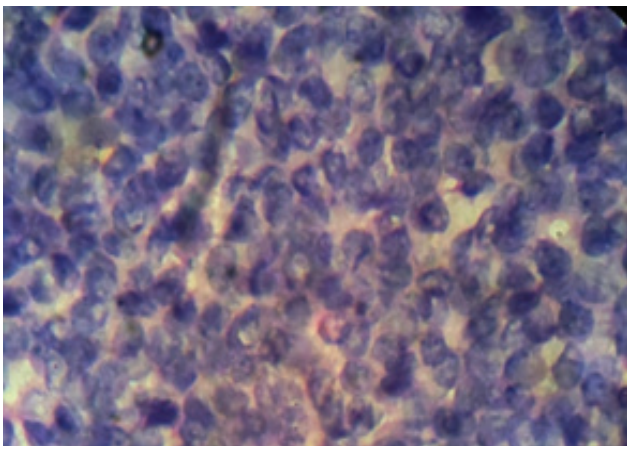

Fig. 3: Showing cells round to ovoid nuclei, scanty cytoplasm and occasional mitotic figure(H&E,100X)

tumors. [4] According to the theory proposed by Motta and Makabe [5] both coelomic epithelium and mesonephros may contribute to the origin of pregranulosa cells. The influence of mesonephros in the origin of the sex cord explains why the sites of extraovarian sex-cord stomal tumors are limited to the broad ligament, retroperitoneum, and adrenals; all of these differentiate close to the mesonephros or the mesonephric duct [5].

The clinical signs and symptoms of extraovarian GCT , based on a study of cases published in literature, are similar to those of ovarian GCT. These symptoms include presence of a mass, irregular vaginal bleeding, postcoital bleeding, and hemiperitoneum. Extraovarian granulosa cell tumor have been seen to develop in retroperitoneum [6] [7] , broad ligament [8], mesentery, omentum, liver, adrenals, and so forth [6].

Several histologically similar tumors need to be differentiated from GCTs - these include undifferentiated carcinoma, small-cell or neuroendocrine carcinoma, endometrial stromal sarcoma, carcinoid, malignant melanoma, intraabdominal desmoplastic small-cell tumor [9] GCTs are characterized by immunopositivity for calretinin, melan-A, inhibin-alpha, and progesterone receptors and by negativity for EMA. In our case, the tumor was positive for inhibin. This along with characteristic histopathological features helped in clinching the diagnosis. Only three previous cases of extraovarian granulose cell tumor have been previously reported from India, to the best of our knowledge.[6][10][11]

Conclusion

It needs to be emphasised that it may be impossible to diagnose these cases in the preoperative work up of the patient and in such cases, it is only after surgery that we arrive at a diagnosis. Extraovarian granulosa cell tumor needs to be kept in mind in a female patient with a retroperitoneal mass. The prognosis for extraovarian sex-cord stromal tumors seems favorable; however, long-term follow-up of these patients needs to be done.

Acknowledgements

Nil

Funding

None

Competing Interests

None

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