Volume-4, Issue-6, June-2015 • ISSN No 2277 - 8160

Research Paper | Medical Science

Unilocular adult type of granulosa cell tumor ovary- a case report.

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ABSTRACT

Granulosa cell tumors are the most frequent sex cord stromal tumors and account for approximately 5% of all primary ovarian tumors. Here we present a case of unilocular granulosa cell tumor in a 27yr old female who presented with acute pain abdomen without any other signs or symptoms. Ultrasonographically a diagnosis of hemorrhagic cyst vs chocolate cyst was. The cystic mass in left ovary measured 14x 12.5 cm. Histopathologically a diagnosis of unilocular adult type granulosa cell tumor was made based on nuclear grooving and Call-Exner bodies. This case had an younger age occurrence, unilocular cystic presentation and absence of features of hyperestrinism or virilisation.

KEYWORDS : unilocular, granulosa cell tumor, ovary.

INTRODUCTION:

Granulosa cell tumors are the most frequent sex cord stromal tumors and account for approximately 5% of all primary ovarian tumors. About 5% occur before puberty and 40% occur in the menopausal age group.[1] These tumors may be oestrogenic, non-functional,or on rare occasions

virulising. Granulosa cell tumors have a predominantly solid cut surface with frequent cystic degeneration and haemorrhage. Unilocular and multilocular cystic granulosa cell tumors have been reported in the literature but are rare.[2,3]

CASE REPORT:

A 27yr old female came to gynaecology OPD of KIMS, Hubli,with history of 4 months amenorrhoea and lower abdominal pain since 5 days. She had no symptoms or signs of hyperestrinisim or virilization.

General physical examination was normal. Abdominal examination revealed a mass consistent with 20-22 weeks pregnancy. The mass was mobile and cystic.

Pelvic ultrasound examination showed a large well defined unilocular midline cyst cranial to uterus extending into abdomen measuring 14.4x16x9.2 cm with diffuse low levels echo within and no evidence of any solid component or calcifications.Ovaries were not adequately visualized. Endometrial thickness was 7mm. A possible diagnosis of chocolate cyst of ovary vs hemorrhagic cyst was made. Lab investigations were within normal limits with CA 125 levels being 24.47U/ml. Patient underwent exploratory laparotomy.

Intraoperatively left ovary showed a haemorrhagic cyst measuring 14x12.5cm and weighed 1.8kg. The right adnexa and other abdominal viscera appeared normal. Left ovarectomy was done.

Grossly, the external surface of cyst wall was smooth and cut surface revealed unilocular cyst with an edematous wall. (Fig. 1:A& B)

Histopathology revealed a unilocular cystic lesion lined by multi- layered granulosa cells with interspersed Call- Exnerbodies(Fig 2 and 3). The neoplastic cells were round to polygonal with moderate to scant cytoplasm and round to oval nuclei with some showing prominent grooving(Fig 4). Occassional mitotics was noted. There was no lymphovascular invasion. A final diagnosis of unilocular adult- type granulosa cell tumor was made.

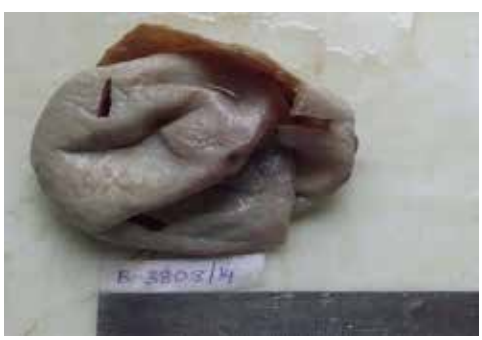

Fig 1(A)

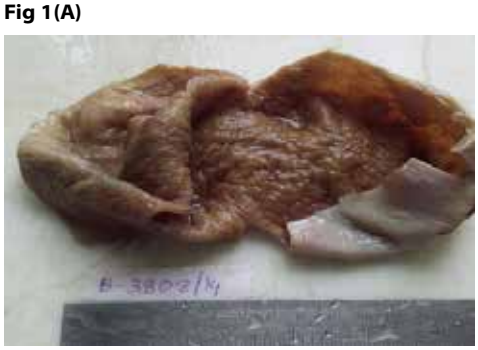

Fig 1(B)

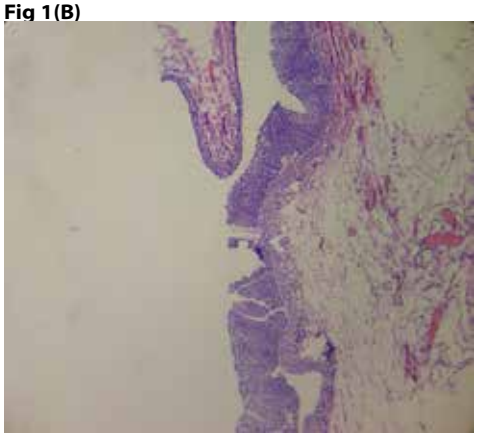

Fig 2(A)

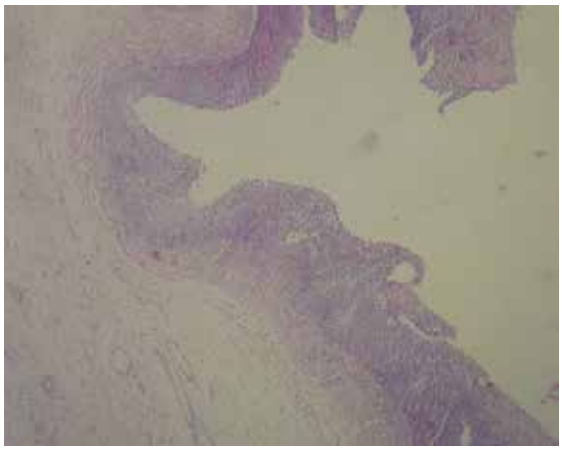

Fig 2(B)

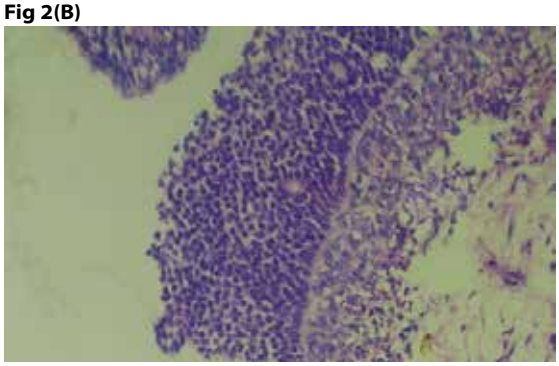

Fig 3(A)

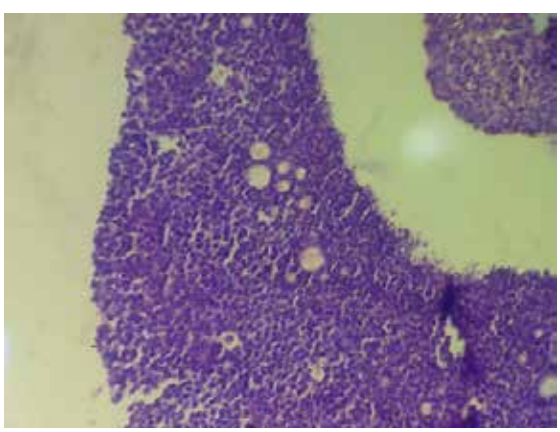

Fig 3(B)

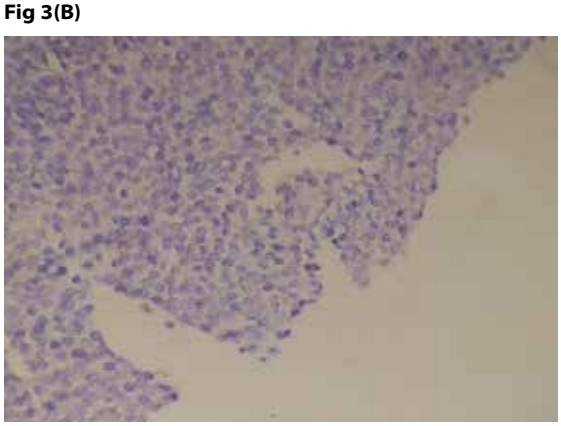

Fig 4.

DISCUSSION:

Sex cord stromal tumors comprise approximately 8% of primary ovarian neoplasms, and among these tumors, the granulosa cell tumor is the most common and accounts for approximately 1.5% of primary ovarian tumors. Histogenesis of sex cord stromal tumors is uncertain; however, at present, the origin is considered to be coelomic epithelium or gonadal sex cord.[2]

There are two well-defined patterns of granulosa cell tumor, the common adult granulosa cell tumor and the less frequent juvenile type, which are based on clinical and histopathological features. The adult granulosa cell tumor accounts for 95% of all ovarian tumors, and most commonly presents in the peri or postmenopausal period with a peak incidence between 50–54 years.[2]

The usual presentations of these tumors are symptoms associated with hyperestrinism leading to isosexualpseudoprecocity in children and metrorrhagia in adults. A large number of androgenic granulosa cell tumors present as unilocular or multilocular cysts.[4,7]

The association with androgen production and the formation of unilocular cystic type of granulosa cell tumor remains an enigma.[5]

Grossly, granulosa cell tumors are usually unilateral and encapsulated with a smooth lobulated outline and predominantly solid or solid and cystic with a yellow to white cut surface. Cystic degeneration and hemorrhage are common. Occasionally, the tumors may resemble a thin-walled unilocular or multilocular cystadenoma.[2]

The histomorphology of adult granulosa cell tumors includes well-differentiated and less well-differentiated types. The well-differentiated group is composed of microfollicular, macrofollicular, trabecular, and insular patterns with the microfollicular pattern being the most common of all subtypes and contains Call-Exner bodies. Diffuse and watered silk or gyriform patterns fall under the less well differentiated group. Sclerotic stroma is a secondary degenerative change commonly seen in granulosa cell tumors, which most likely arises due to ischemia.[5]Differential diagnosis for granulosa cell tumors include low-grade stromal sarcomas, small cell carcinomas, and carcinoid tumors on low power magnification. These tumors lack nuclear grooving, are more hyperchromatic, and often contain more mitotic figures than typical granulosa cell tumors. The nuclear appearance, mitotic rate, and presence of Call-Exner bodies have a diagnostic value in differentiating granulosa cell tumors from other malignant tumors.

Elevated levels of alpha inhibin and/or Müllerian-inhibiting substance (MIS) or anti-Müllerian hormone are useful and specific markers for early diagnosis and follow-up of granulosa cell tumors.

Here,the diagnosis of unilocular cystic granulosa cell tumors is based on the histological examination with a monotonous population of round to oval cells with Call-Exner bodies, nuclear grooves, low mitotic activity

Granulosa cell tumors are slow growing tumors, and late recurrence is a common feature. Clinicopathologic prognostic markers include the tumor stage as well as tumor size and rupture, of which staging is the most important prognostic factor.[6]

The 5-year survival rate for stage 1 disease ranges from 75% –95%. Presence of tumors greater than 10–15 cm in diameter carries a poor prognosis independent of stage. A mitotic index of more than or equal to 10 mitotic figures per high power field (×10) has a poor prognosis. Other histopathological variables used to determine prognosis include p53 status, histological pattern, disease stage, mitotic index, and lymphovascular invasion. Of these factors, mitotic index and lymphovascular invasion were the most important and are independent factors that determine prognosis.[2]Life-long follow-up with clinical examinations and measurement of tumor markers such as inhibin B are recommended.

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