

Juvenile granulosa cell tumor in a 35-year-old pregnant woman: A rare case report

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ABSTRACT

Juvenile granulosa cell tumors (JGCTs) account for 5% of GCT and 3% are diagnosed after 30 years of age. JGCT is an uncommon neoplasm in pregnancy. About 90% are stage I at diagnosis but advanced-stage tumors are aggressive often resulting in recurrence or death within 3 years of diagnosis. We present a case of JGCT in a pregnant woman of 35 years old. The patient complained of sudden onset pain in her second trimester. Ultrasound whole abdomen was done and a malignant lesion was detected. Subsequently, the left ovarian cystectomy was performed and a diagnosis of JGCT was given which was confirmed by histopathology and immunohistochemistry.

Key words: Granulosa cell tumors, Juvenile granulosa cell tumors, Pregnancy

Juvenile granulosa cell tumors (JGCTs) are rare, accounting for 5% of all GCTs, which in total, make up 5% of all gynecologic malignancies [1]. Approximately, 80% of cases are diagnosed during the first two decades of life. The average age of diagnosis of JGCTs is 13 years old, and only 3% are diagnosed in women over 30 years [2]. Only 10% of GCTs present during pregnancy, accounting for <1% of ovarian neoplasms diagnosed in pregnancy [3,4].

This report describes a case of JGCT in a pregnant woman at 14 weeks' gestation, with the rapid growth of the mass and presenting with pain abdomen. We are presenting this case as JGCT is an uncommon neoplasm in pregnancy and only a few cases have been reported in the literature to date.

CASE REPORT

A 35-year-old pregnant woman came to our hospital at 14 weeks of gestation complaining of sudden onset pain in the abdomen and lower back region on the left side for 2 days. She had no significant medical or surgical history.

The general examination was done and was normal. The vitals of the patient were within normal range. Ultrasound whole abdomen was done and an adnexal mass, 90 cm in diameter was noted. Blood tests and serum markers such as CA 125, CEA, CA 19.9, AFP, LDH, and beta HCG were done and were found to be within normal limits.

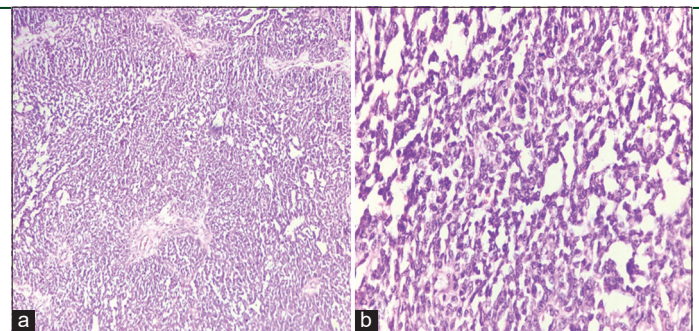



Figure 1: (a) Photomicrograph showing sheets and trabeculae of granulosa cells surrounding follicle-like cystic spaces in low power view; (b) Photomicrograph showing round to oval cells with hyperchromatic vesicular nuclei, small to prominent nucleoli and irregular nuclear contours in high power view. Occasional nuclear grooves were noted. Numerous mitotic figures were noted

The case was then planned for surgery. Abdominal left ovarian tumor resection was performed. The specimen was received in our department in 10% formalin. Grossly, the tumor was of size 13×12×5 cm with the ruptured capsule. The cut surface was solid and greyish-white with focal papillary and hemorrhagic areas. Section was made of 3–5 micron thickness and stained with H&E.

Histopathological examination showed round to oval cells with hyperchromatic vesicular nuclei, small to prominent nucleoli, and irregular nuclear contours in high power view (Fig. 1). Occasional nuclear grooves and numerous mitotic figures were noted (Fig. 1b). Immunohistochemistry for markers such as WT1, panCK, Calretinin, and CK 7 was done and showed strong diffuse nuclear positivity for WT1 (Fig. 2a), patchy positivity for pan CK (Fig. 2b), and focal positivity for calretinin

Access this article online	
Received - 17 November 2023 Initial Review - 30 November 2023 Accepted - 06 January 2024	Quick Response code 
DOI: 10.32677/ijcr.v10i2.4357	

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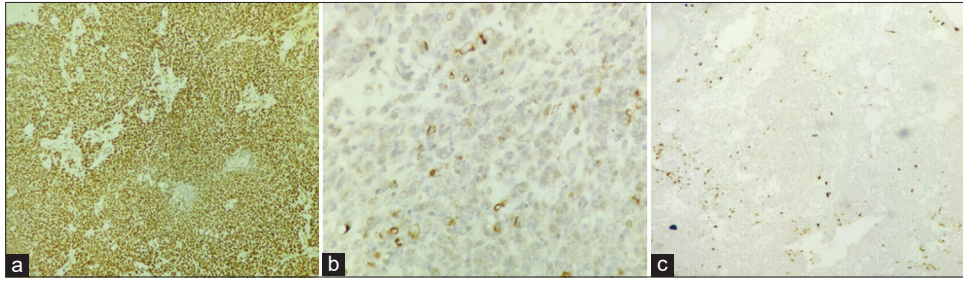


Figure 2: Photomicrograph showing strong (a) diffuse nuclear positivity for WT1; (b) patchy positivity for pan CK; (c) focal positivity for Calretinin

(Fig. 2c). CK 7 was negative. Thus, histopathological findings and immunohistochemical studies led to the diagnosis of JGCT.

The patient again had a recurrence 3 months later for which she underwent a left ovariectomy. Histopathological examination of the left ovariectomy specimen showed JGCT. She delivered a healthy baby at term 2 months later by cesarean section. She had heavy episodes of vaginal bleeding at the time of delivery. Total abdominal hysterectomy and salpingo-oophorectomy were performed. On HPE, the patient was again positive for JGCT. The patient developed ascites a few weeks later. The patient was advised adjuvant chemotherapy and is under follow-up.

DISCUSSION

GCTs are part of the sex cord-stromal tumors that occur with a rare incidence rate that makes up 2–5% of all ovarian malignancies. These tumors are classified into 2 distinct subtypes: adult GCT (AGCT) and juvenile GCT (JGCT), with the juvenile type constituting only 5% of GCTs and occurring predominantly in women under the age of 30 years [5]. GCT is a rare malignancy, and its detection in pregnancy is even more so. The incidence of ovarian carcinoma diagnosed during pregnancy varies from 0.0179 to 0.11/1000 pregnancies [6]. In the largest study of JGCTs to date performed by Young *et al.* in 1984, only 4 (3.2%) of 125 patients were over 30 years old [4]. In our case also, the patient was of 35 years old.

The most common presenting symptoms of both AGCT and JGCT are abdominal pain and increasing abdominal girth [2]. In our case also, the patient presented with acute onset abdominal pain and lower back pain. The use of imaging modalities cannot adequately distinguish JGCT from other ovarian neoplasms due to the absence of any characteristic features. Therefore, evaluation using histology and immunochemical assays can help provide a confirmatory diagnosis [7].

Histologically, AGCTs are characterized by the presence of Call–Exner bodies, which are small, eosinophilic fluid-filled spaces between granulosa cells resembling ovarian follicles, and “coffee-bean” nuclei, which are grooved, pale nuclei with a low mitotic rate. In contrast, JGCTs have rare Call–Exner bodies and cells contain abundant eosinophilic or vacuolated cytoplasm with non-grooved, larger, hyperchromatic nuclei with mild-to-severe nuclear atypia and higher mitotic rate than adult types [8]. Pathology of this patient’s tumor lacked Call–Exner bodies and

nuclei were hyperchromatic without nuclear grooves, consistent with diagnosis of JGCT. Immunohistochemistry for markers such as WT1, panCK, Calretinin, and CK 7 was done and showed strong diffuse nuclear positivity for WT1, patchy positivity for pan CK, and focal positivity for Calretinin. CK 7 was negative.

In the case of GCTs, studies have shown that during pregnancy there is a disorderly arrangement of tumor cells that results in the absence of recognizable differentiation in many areas, as well as, prominent edema and large numbers of luteinized cells. These patterns are not frequently observed in juvenile GCTs of non-pregnant subjects [6].

Treatment strategies include conservative management until after delivery or surgical management. Conservative management has been suggested in asymptomatic adnexal masses <6 cm but can result in adverse pregnancy outcomes including rupture, torsion, or hemorrhage. In patients with persistent adnexal masses >6 cm, surgical removal should be delayed until the second trimester if possible due to the theoretical risk of spontaneous abortion in the first trimester with anesthesia [4].

Adjuvant chemotherapy is recommended for patients with stage 1C disease or higher or in cases with a high mitotic index greater than 20/10 high power fields. Ten adequate follow-ups after initial treatment are crucial in these cases. Routine follow-up should be done at 2–3 month intervals for the first 2 years followed by 4–6-month intervals over the next 3 years. Then there should be annual follow-ups for another 5 years. Any evidence of recurrence should be followed up with imaging of the abdomen and pelvis where the physician should look for recurrent tumors [5].

CONCLUSION

JGCT is an uncommon neoplasm in pregnancy. About 90% are stage I at diagnosis, and confined to one ovary. However, advanced-stage tumors are aggressive often resulting in recurrence or death within 3 years of diagnosis. The mainstay of treatment is surgery with the addition of adjuvant chemotherapy for advanced-stage disease. We report this interesting case that can be especially dangerous in pregnancy since it often goes unnoticed.

ACKNOWLEDGMENT

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Sharma JD, Kalita U, Deka M, Bharadwaj B, Beso AP. Juvenile granulosa cell tumor in a 35-year-old pregnant woman: A rare case report. *Indian J Case Reports*. 2024;10(2):38-40.