Adult Granulosa Cell Tumor of the Ovary Diagnosed after Laparoscopic Salpingoophorectomy

Ansari SN, Shrestha E, Kayastha P, Bista KB

Department of Gyneconcology

National Academy of Medical Sciences

Kathmandu, Nepal.

Corresponding Author

Shaheen Naaz Ansari

Fellow in Gyneconcology,

Department of Gyneconcology,

National Academy of Medical Sciences,

Kathmandu, Nepal.

E-mail: drshaheenansari21@gmail.com

Citation

Ansari SN, Shrestha E, Kayastha P, Bista KB. Adult Granulosa Cell Tumor of the Ovary Diagnosed after Laparoscopic Salpingoophorectomy. *Kathmandu Univ Med J.* 2025; 91(3): 392-4.

ABSTRACT

Ovarian malignancies of uncommon variety are seen in 10% of cases of all ovarian tumors. Approximately 5% of such cases are granulosa cell tumor of ovary (GCT). In adolecents, the adult type Granulosa cell tumor (AGCT) is rare and the major concern is fertility preservation. Prognosis of the adult type granulosa cell tumor is favorable after surgery, as most cases present at early stages.

We report a case of an 18-year girl who presented to our center with histopathological report of the adult type granulosa cell tumor. She underwent laparoscopic left salpingoophorectomy at another center for a 28.5 X 22.5 cm right adenexal mass. The patient denied completion surgery at our center and remained on follow-up. Patient is doing fine on follow-up to date.

In young girls who present with complex adenexal mass, a strong suspicion of rare ovarian malignancies like granulosa cell tumor should be considered. For cases with incomplete surgery but fertility concern granulosa cell tumor should be monitored vigilantly for recurrence.

KEY WORDS

Adenexal mass, Adolescence, Fertility preservation, Granulosa cell tumor

INTRODUCTION

Granulosa cell tumors (GCT) are rare malignancies of ovary with incidence of 0.5-1.5/100000 per women per year.[1] It is the most common type of sex cord stromal tumor constituting about 70% of cases.[2] Up to 90% of the cases present in Stage I.[3] Primary treatment for early stage granulosa cell tumor is comprehensive staging with unilateral salpingooophorectomy.

We report a case of an 18-year adolescent girl who presented to us with histopathological report of adult type granulosa cell tumor following left salphingoophorectomy at another institute, and at our institute, she denied repeat laparotomy for complete staging procedure.

An incompletely staged surgery for a granulosa cell tumor in an adolescence who has fertility concern but denies further surgery needs to be managed with individualized plan.

CASE REPORT

An 18 years girl presented to our hospital with histopathological report of adult type granulosa cell tumor 2 months after left laparoscopic salpingoophorectomy performed at another center. Her menarche occurred at 14 years. Her preoperative CECT showed left ovarian large unilocular cyst of 28.5 X 22.5 cm and right ovary of 33X24 mm. Serum Inhibin B was 13.23 pg/ml and LDH was 744 U/L, all other tumor markers were normal. Her intraoperative and postoperative event was unremarkable. At our center, a post-operative CECT showed no evidence of residual or recurrent tumor, no significant lymphadenopathy, no ascites or peritoneal deposits. On examination, her general condition was fair. Abdomen was soft and non-tender, wound was healthy. Her reports were evaluated and histopathological slides were reviewed at our center which showed adult type granulosa cell tumor confined to ovary,

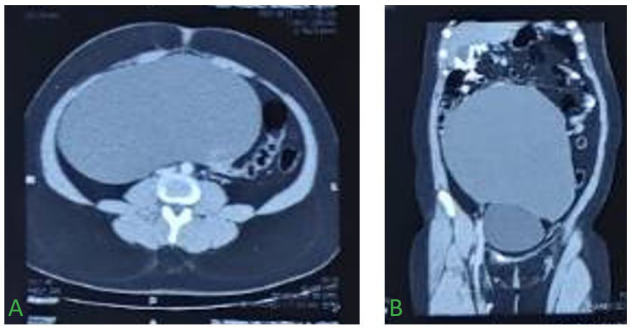

Figure 1. CECT showing left ovarian low attenuation mass of 28.5 X 22.5 cm. A. Coronal view. B. Sagittal view.

cyst wall showed sloughed off lining. Mitotic activity was 4/5 hpf. Completion surgery for staging was planned but the patient denied repeat surgery due to fertility concern. As per the discussion of the local tumor board, she was counselled for completion surgery as first option and chemotherapy as other option. She denied both and lost to follow-up. She presented after 5 months with amenorrhea for 5 months. CECT abdomen and pelvis was done which was normal. Inhibin was 44.67 pg/ml. She opted to remain on follow-up and go for surgery if indicated during follow-up. At 6 weeks ultrasound pelvis and tumor markers were normal. Thereafter, follow-up 3 monthly with ultrasound abdomen and pelvis and tumor markers was done and the reports remained within normal range. She is asymptomatic with no features of recurrence until 3 years and 10 months of follow-up period now.

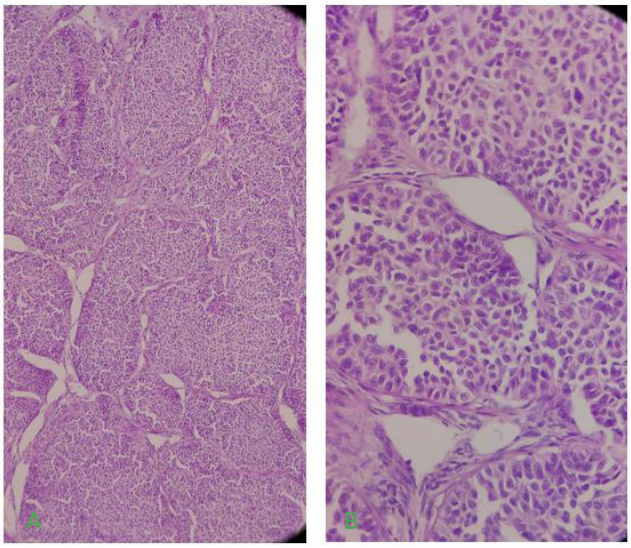

Figure 2. Histological slides of showing A. proliferation of granulosa cells arranged in sheets. B. Mitotic figures are noted.

DISCUSSION

Granulosa cell tumor arises from granulosa cells of the ovary. Granulosa cells produce estradiol, AMH inhibin A and inhibin B. Excess estradiol produced by the tumor results in menstrual symptoms like irregular menstruation and heavy flow. Estradiol also increases risk of endometrial hyperplasia and breast cancer.[5,6]

GCT are large ovarian tumors with average size of 10-15 cm. The size of tumor in this case was larger. GCT is of two types, adult type, which constitutes 95% cases and 5% is juvenile type depending on the natural history and histologic features.[7] The peak incidence of AGCT lies between 50 and 55 years, while about 1% of AGCT cases are observed in adolescence girls.[4] Ultrasound features of GCTs are large multilocular solid masses or solid tumors with heterogeneous echogenicity. Other radiological features are irregular, thick walls and septa, papillary projections, solid and echogenic foci.[8] On CECT granulosa cell tumor appear as well defined thick walled unilocular, low attenuation masses, multilocular cysts or heterogeneously solid masses. Immunohistochemistry positivity of granulosa cells is seen with inhibin-α, WT-1, and CD56.

In young patients of reproductive age, a fertility-sparing approach to management should be considered and such patients must be regularly monitored for relapse and recurrence. The most important indicator of prognosis and recurrence is surgical staging.[9] A complete staging for granulosa cell tumor includes peritoneal washings, careful examination of the abdominal cavity, salpingooophorectomy, peritoneal biopsies, omentectomy and endometrial biopsy. Preservation of the uterus and contralateral ovary is safe with excellent prognosis and low recurrence in macroscopic stage IA granulosa cell tumor in young patients.[4,10]

A completion surgery with compreshensive staging is needed for cases that are incompletely staged at initial surgery to estimate the actual stage of the disease to guide need of further adjuvant management. Stage IA granulosa cell tumors have an excellent prognosis after complete surgical staging with salpingoophorectomy alone without adjuvant therapy. Adjuvant chemotherapy is given in stage IC2-1C3 of AGCT and more advanced disease or for irresectable tumors.[11] In very young girls with incompletely staged surgery as in this case a completion surgery following family completion on patient's wish or when indicated on surveillance can be a good approach.

Follow up of conservatively managed cases involves estimation of tumor markers AMH and Inhibin B and pelvic scan along with MRI imaging or CECT if indicated. Biomarkers AMH and inhibin B accurately correlate with disease status.[4] Small case series have reported low recurrence rate of 5.4% in Stage I increasing to 40% in stage IV disease.[12] One-third of patients typically relapse within 4 to 7 years following initial diagnosis. Recurrence may sometimes be very late after tumor resection, and can be as late as 20-30 years.[13] Recurrent disease is generally more aggressive in nature and risk of death is higher as compared to initial disease.

Young girls diagnosed with GCT at early stage have good prognosis with fertility sparing surgery but should be monitored closely for relapse or recurrence.

REFERENCES

1. Kottarathil VD, Antony MA, Nair IR, Pavithran K. Recent advances in granulosa cell tumor ovary: a review. *Indian J Surg Oncol.* 2013 Mar;4(1):37-47. doi: 10.1007/s13193-012-0201-z. Epub 2012 Dec 7. PMID: 24426698; PMCID: PMC3578540.
2. Hashemipour M, Moaddab MH, Nazem M, Mahzouni P, Salek M. Granulosa cell tumor in a six-year-old girl presented as precocious puberty. *J Res Med Sci.* 2010 Jul;15(4):240-2. PMID: 21526089; PMCID: PMC3082815.
3. Lee IH, Choi CH, Hong DG, Song JY, Kim YJ, Kim KT, et al. Clinicopathologic characteristics of granulosa cell tumors of the ovary: a multicenter retrospective study. *J Gynecol Oncol.* 2011 Sep;22(3):188-95. doi: 10.3802/jgo.2011.22.3.188. Epub 2011 Sep 28. PMID: 21998762; PMCID: PMC3188718.
4. Kim YS, Lee JH. A case report of ovarian granulosa cell tumor in patient with polycystic ovarian syndrome. *Medicine* (Baltimore). 2021 Dec 17;100(50):e28261. doi: 10.1097/MD.0000000000028261. PMID: 34918698; PMCID: PMC10545264.
5. Boyce EA, Costaggini I, Vitonis A, Feltmate C, Muto M, Berkowitz R, et al. The epidemiology of ovarian granulosa cell tumors: a case-control study. *Gynecol Oncol.* 2009 Nov;115(2):221-5. doi: 10.1016/j.ygyno.2009.06.040. Epub 2009 Aug 7. PMID: 19664811.
6. Hammer A, Lauszus FF, Petersen AC. Ovarian granulosa cell tumor and increased risk of breast cancer. *Acta Obstet Gynecol Scand.* 2013 Dec;92(12):1422-5. doi: 10.1111/aogs.12252. Epub 2013 Oct 28. PMID: 24283356.
7. Young RH. Sex cord-stromal, steroid cell, and other ovarian tumors with endocrine, paraendocrine, and paraneoplastic manifestations. InBlaustein's pathology of the female genital tract 2011 (pp. 785-846). Springer, Boston, MA.
8. Kim JA, Chun YK, Moon MH, Lee YH, Cho HC, Lee MS, et al. High-Resolution Sonographic Findings of Ovarian Granulosa Cell Tumors: Correlation With Pathologic Findings. *J Ultrasound Med*. 2010 Feb;29(2):187-93.
9. Pectasides D, Pectasides E, Psyrri A. Granulosa cell tumor of the ovary. *Cancer Treat Rev.* 2008 Feb;34(1):1-12. doi: 10.1016/j.ctrv.2007.08.007. Epub 2007 Oct 22. PMID: 17945423.
10. Levin G, Zigron R, Haj-Yahya R, Matan LS, Rottenstreich A. Granulosa cell tumor of ovary: A systematic review of recent evidence. *Eur J Obstet Gynecol Reprod Biol.* 2018 Jun;225:57-61. doi: 10.1016/j.ejogrb.2018.04.002. Epub 2018 Apr 11. PMID: 29665458.
11. Ray-Coquard I, Morice P, Lorusso D, Prat J, Oaknin A, Pautier P, et al. ESMO Guidelines Committee. Non-epithelial ovarian cancer: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol*. 2018 Oct 1;29(Suppl 4):iv1-iv18. doi: 10.1093/annonc/mdy001. PMID: 29697741.
12. Färkkilä A, Haltia UM, Tapper J, McConechy MK, Huntsman DG, Heikinheimo M. Pathogenesis and treatment of adult-type granulosa cell tumor of the ovary. *Ann Med.* 2017 Aug;49(5):435-47. doi: 10.1080/07853890.2017.1294760. Epub 2017 Mar 6. PMID: 28276867.
13. Park H, Goodman CP, Raymond SL, Sundin A, Khan FA, Radulescu A. A 12-Year-Old Girl with Juvenile Granulosa Cell Tumor of the Ovary, Presenting with Adolescent Hyperprolactinemia, Galactorrhea, and Amenorrhea. *Am J Case Rep*. 2023 Jan 17;24:e938249. doi: 10.12659/AJCR.938249. PMID: 36647328; PMCID: PMC9867898.