Journal of Pediatric Surgery Case Reports 76 (2022) 102093

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.elsevier.com/locate/epsc

Torsion of juvenile granulosa cell ovarian tumor

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ARTICLE INFO

Keywords:
Juvenile granulosa cell tumor
Alpha-inhibin
Immunohistochemistry
Ovarian sex cord-stromal tumors

ABSTRACT

Juvenile granulosa cell tumor (JGCT), which is one of the sex cord-stromal tumors of the ovary, is a rare malignancy of childhood. Juvenile type is a rare form that accounts for 5% of granulosa cell tumors. Isosexual precocious puberty is the most common presentation in prepubertal girls with juvenile granulosa cell tumors. Less frequently, they present with the complaint of a mass in the abdomen.

We present an 8-year-old girl with a juvenile granulosa cell tumor who presented with sudden onset of abdominal pain and enlargement of the abdomen. She underwent exploratory laparotomy with right salpingo-oophorectomy and a complete mass resection was performed. Pathologic staging of the mass, which was diagnosed as juvenile granulosa cell tumor of the ovary, was determined as pT1c3, and our patient received four cycles of chemotherapy.

1. Introduction

Ovarian tumors are responsible for approximately 1% of childhood cancers [1]. Tumors of ovarian origin are classified as epithelial tumors, sex cord-stromal tumors and germ cell tumors [2,3]. Sex cord-stromal tumors of the ovary are rare and constitute approximately 10–15% of ovarian tumors in girls [4]. Granulosa cell tumors (GCTs) are thought to arise from estrogen-producing granulosa cells that surround the germinal cells in the ovarian follicles. It has two types, adult and juvenile, with different histological and clinical features. JGCTs are rare tumors seen in all premenarche girls and young women. JGCT is a rare ovarian tumor that is the most common ovarian sex cord stromal tumor in the pediatric age group. JGCTs are often associated with precocious puberty due to excessive estrogen production [4–7].

2. Case report

We present an 8-year-old girl with rapid onset abdominal pain followed by abdominal distension. There was no history of associated vomiting, but nausea. On abdominal examination, the mass occupying the right lower abdomen and extending to the umbilicus and middle was palpated. The patient didn't have evidence of breast and pubic hair development. Genital examination revealed a morphologically normal female with normal hymenal opening. Other aspects of the patient's physical examination were also normal.

Laboratory and imaging studies were performed. Electrolytes, CBC, liver function tests, and LDH were normal. Preoperative tumor markers, including AFP, β-hCG, CA 125, CEA, and CA 19.9 were negative. Inhibin levels could not be measured because tests were not available at our laboratory. A pelvic ultrasound revealed a mass measuring 12.0 × 11.0 × 7.2 cm. There were hyperechogenities of

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https://doi.org/10.1016/j.epsc.2021.102093
Received 8 October 2021; Received in revised form 17 October 2021; Accepted 21 October 2021
Available online 1 November 2021

solid components and anechoic-hypoechoic areas of cystic components within the lesion. Magnetic Resonance Imaging (MRI) confirmed the presence of a mass originating from the right adnexa (Fig. 1a–d). In conventional sequences, apart from solid-cystic components supporting US, areas of restriction in diffusion-weighted imaging (DWI) and heterogeneous contrast enhancements especially in solid components in the lesion were observed in post-contrast images. MRI also showed that the lesion caused grade-2 hydronephrosis by compressing the right ureter. There was also a minimal localized intraperitoneal fluid signal in the immediate inferior side of the lesion. On computed tomography (Fig. 1e and f), the heterogeneous smoothly contour lesion displaces the bowel loops (on axial image-e) and extends superiorly to the level of the L3 vertebra and inferiorly to the level of the bladder apex, and the bladder is slightly compressed by the lesion (on sagittal image-f).

Laparotomy performed with an infraumbilical midline insicion revealed involvement and 360° torsion of the right ovary (Fig. 2a and b). During exploration, samples were taken from intraabdominal serous fluid for cytology. After the cyst had been aspirated and decompressed, it was pulled out through the incision, the torsion was untwisted by rotating the tumor, and a right salpingo-oophorectomy was completed with total mass resection. There were no intraoperative and postoperative complication.

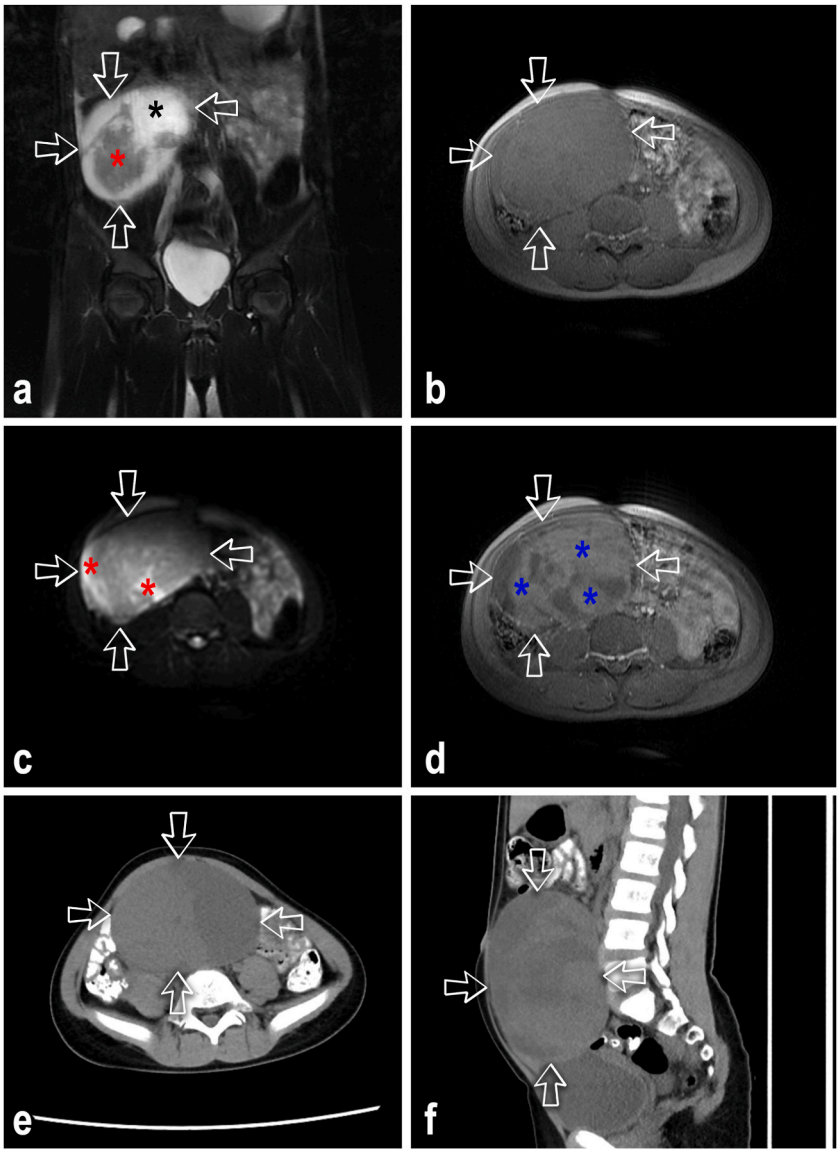

Fig. 1. (MR images of the lesion (open arrows)): a. In the coronal T2 fat supressed image, a mass lesion with a heterogeneous signal in the right side, deplasing abdominal wall anteriorly. In the lesion high signal areas due to cystic-necrotic (black asterix) components, and intermediate intensity areas (red asterix) indicate solid components were seen. b, c and d images of axial sections; isointense appearance with muscle tissue in T1 weighted image (b), the restriction areas (red asterixis) showing hypercellularity in the DWI sequence (c) and contrast enhancement of the solid components (blue asterixis) in postcontrast examination (d) are shown. Axial (e) and sagittal (f) CT images of the mass lesion. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

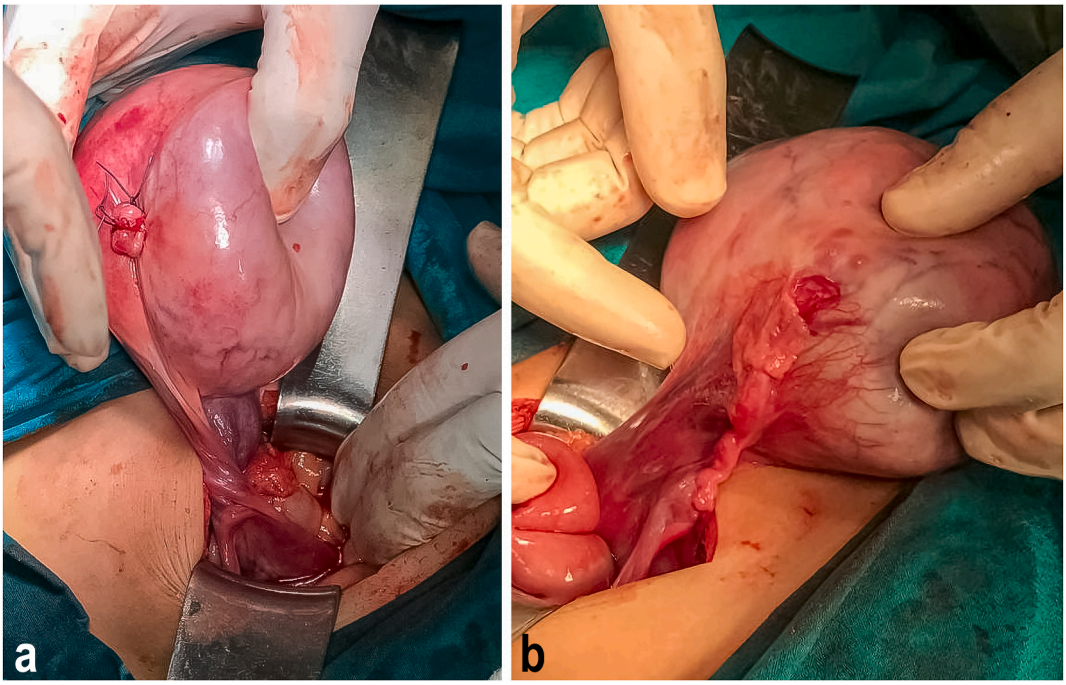

Fig. 2. Juvenile granulosa cell tumor of the right ovary in 8 year-old girl, (a)a twisted ovarian pedicle (arrow), (b) detorsion completed before salpingo-oophorectomy.

Intraoperative and postoperative complication were not encountered.

Macroscopic evaluation revealed an 11.0 × 10.0 × 7.0 cm diameter ovarian mass with a 4.0 × 0.3 cm fallopian tube. The surface of ovarian mass was smooth and intact. A tumoral formation with a 9 cm diameter revealing irregular borders, hemorrhagic, solid, and cystic areas was observed on the cross-sectional surface of the specimen. Due to the tumor mass, normal ovarian parenchyma could be found only in focal areas.

Microscopically, the tumor consisted of follicular pattern with variable size (Fig. 3a), solid (Fig. 3b) and trabecular areas. Tumor cells were small to medium-sized cells with hyperchromatic nuclei and scanty cytoplasm (Fig. 3c). Hemorrhage and focal necrosis areas were observed in non-tumor areas. Immunohistochemical study of the specimens showed positive staining with alpha-inhibin (Fig. 3d) and WT1 in tumor cells, while no staining with TTF1, CD45, CD30, CK7, CK20, PLAP, OCT3/4 was detected. Histopathological diagnosis was reported as a granulosa cell tumor of the right ovary. No tumor was seen in the sections from the fallopian tube and omentum. The pathological stage of the case, in which tumor cells were observed in the cytological examination of the peritoneal washing fluid (Fig. 3e), was accepted as pT1c3.

Our patient received four courses of cisplatin, etoposide, and bleomycin (BEP), and achieved complete clinical remission after initial surgery and adjuvant chemotherapy. No recurrence was observed in our patient with stage Ic disease during the 28-month follow-up.

3. Discussion

JGCT is the most common pediatric ovarian sex cord stromal tumor [5]. In premenarchal girls, JGCTs present often with precocious puberty due to systemic hormonal effects of estrogen overproduction from tumor cells [6,7]. Increasing abdominal girth with a palpable mass and acute abdomen owing to ovarian torsion are other presenting symptoms or signs of JGCTs as we outline in our case report [8,9]. The majority of JGCTs are stage I by FIGO staging at diagnosis and are treated with surgical resection alone, and the result of these tumors is usually favorable, but in stage Ic patients the situation is not as clear as in stage Ia or Ib tumors [10,11]). FIGO stage Ic tumors, which capsule ruptured, tumor on ovarian surface or positive malignant cells in the ascites or positive peritoneal washing, may require postoperative chemotherapy.

Laparoscopic surgery has become the gold standard in the treatment of ovarian cystic tumors in which malignancy can be excluded before surgery. However, laparoscopic ovarian tumor resection is limited because it increases the tumor grade to Stage IC due to intraoperative ovarian cyst rupture and spreading of its contents to the abdomen [12,13]. We preferred laparotomy due to malignancy and torsion suspicion, and the infraumbilical midline incision allowed us to make adequate exploration and comfortable access to the mass. Therefore, attention should be paid to FIGO staging criteria and peritoneal fluid sampling during the operation, and attention should be paid to the identification of stage Ic patients.

Fertility-sparing surgery is preferred in children, adolescents, and women when fertility preservation is desired. There is no need for hysterectomy in stage I tumors as long as the contralateral tube and ovary and the uterus are unaffected, unilateral salpingo-oophorectomy with examination of the contralateral ovary is likely sufficient [14–16]. The negative preoperative tumor markers in

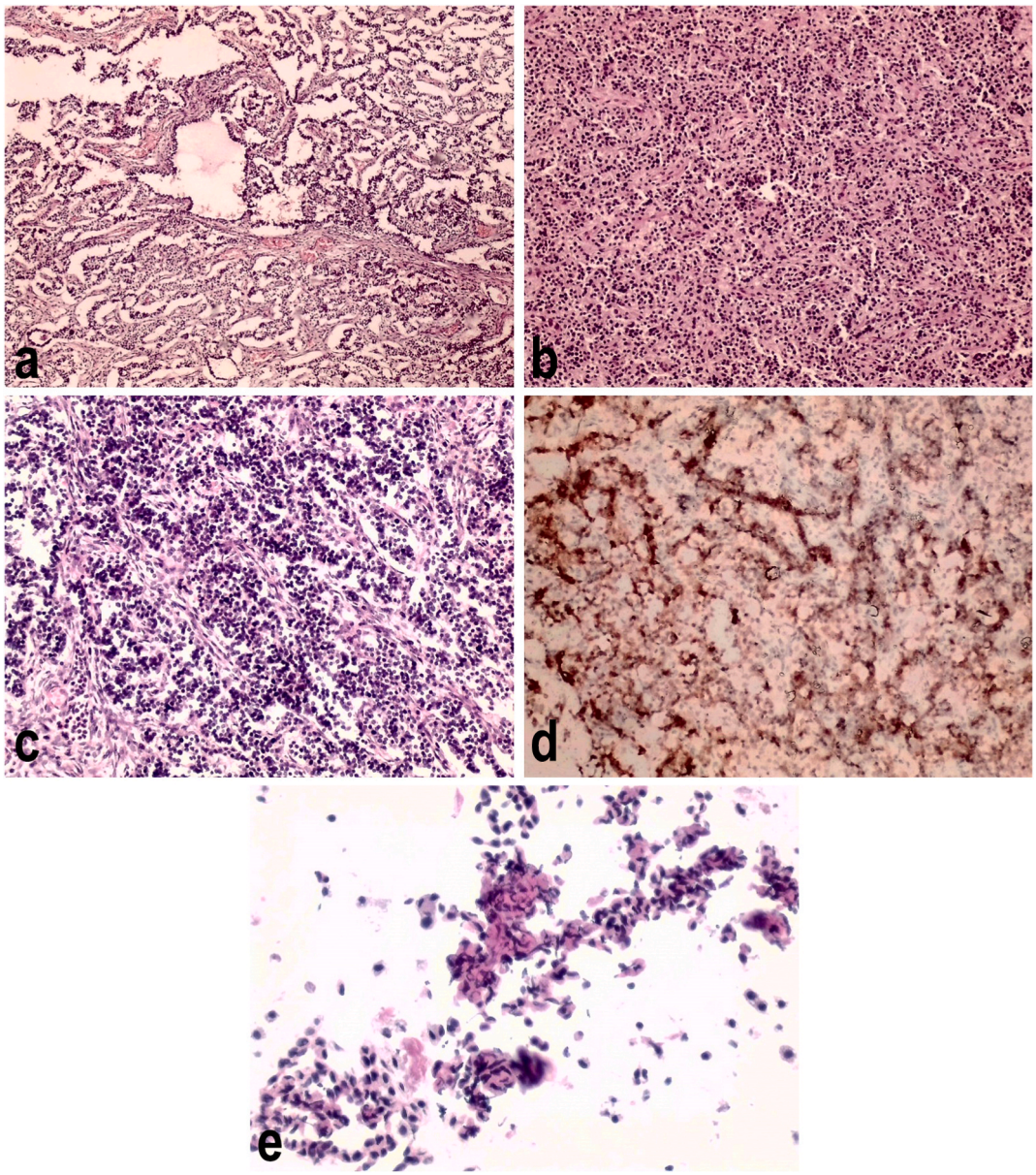

Fig. 3. Variable sized follicular pattern (a) (HE x 40), solid areas (b) (HE x 100). Tumor cells with hyperchromatic nuclei (c) (HE x 100) and inhibin positivity (d) (Alpha-inhibin x 100). Tumor cells in cell block section from peritoneal wash fluid (e) (HE x 200).

our patient did not rule out malignancy. Although there was no evidence of early puberty, which is a possible sign of malignancy in our case, the presence of a large mass with a diameter of 12 cm with cystic and solid components suggested that it was associated with malignancy [17,18].

4. Conclusions

JGCT is the most common pediatric ovarian sex cordstromal tumor. Fertility-sparing surgery is the treatment of choice for ovarian juvenile granulosa cell tumor and the overall prognosis is good. Fertility-sparing surgery may be preferred in reproductive age, if possible.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] Breen JL, Maxson WS. Ovarian tumors in children and adolescents. Clin Obstet Gynecol 1977;20(3):607–23. https://doi.org/10.1097/00003081-197709000-00010.

[2] Chen VW, Ruiz B, Killeen JL, Coté TR, Wu XC, Correa CN. Pathology and classification of ovarian tumors. Cancer 2003;97(10 Suppl):2631–42. https://doi.org/10.1002/cncr.11345.

[3] Kurman RJ, Carcangiu ML, Herrington CS, et al. WHO classification of tumours of female reproductive organs. Lyon: IARC Press; 2014.

[4] Schultz KA, Schneider DT, Pashankar F, Ross J, Frazier L. Management of ovarian and testicular sex cord-stromal tumors in children and adolescents. J Pediatr Hematol Oncol 2012;34(Suppl 2):S55–63. https://doi.org/10.1097/MPH.0b013e31824e3867.

[5] Kottarathil VD, Antony MA, Nair IR, Pavithran K. Recent advances in granulosa cell tumor ovary: a review. Indian J Surg Oncol 2013;4(1):37–47. https://doi.org/10.1007/s13193-012-0201-z.

[6] Sivasankaran S, Itam P, Ayensu-Coker L, Sanchez J, Egler RA, Anderson ML, et al. Juvenile granulosa cell ovarian tumor: a case report and review of literature. J Pediatr Adolesc Gynecol 2009;22(5):e114–7. https://doi.org/10.1016/j.jpag.2008.08.001.

[7] 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;15(4):240–2.

[8] Oltmann SC, Fischer A, Barber R, Huang R, Hicks B, Garcia N. Pediatric ovarian malignancy presenting as ovarian torsion: incidence and relevance. J Pediatr Surg 2010;45(1):135–9. https://doi.org/10.1016/j.jpedsurg.2009.10.021.

[9] Nikumbh DB, Suryawanshi KH, Chitale AM, Pagare P, Surana A. Ovarian juvenile granulosa cell tumour in childhood: uncommon gynecological malignancy. J Clin Diagn Res 2014;8(10):FL01–2. https://doi.org/10.7860/JCDR/2014/9510.4935.

[10] Calaminus G, Wessalowski R, Harms D, Göbel U. Juvenile granulosa cell tumors of the ovary in children and adolescents: results from 33 patients registered in a prospective cooperative study. Gynecol Oncol 1997;65(3):447–52. https://doi.org/10.1006/gyno.1997.4695.

[11] Prat J, FIGO Committee on Gynecologic Oncology. Staging classification for cancer of the ovary, fallopian tube, and peritoneum. Int J Gynaecol Obstet 2014;124(1):1–5. https://doi.org/10.1016/j.ijgo.2013.10.001.

[12] Oue T, Uehara S, Sasaki T, et al. Treatment and ovarian preservation in children with ovarian tumors. J Pediatr Surg 2015;50(12):2116–8. https://doi.org/10.1016/j.jpedsurg.2015.08.036.

[13] Schultz KA, Harris AK, Schneider DT, et al. Ovarian sex cord-stromal tumors. J Oncol Pract 2016;12(10):940–6. https://doi.org/10.1200/JOP.2016.016261.

[14] Abdel-Hady el S, Abdel-Hady Hemida R, Gamal A, El-Shamey M. Fertility sparing surgery for ovarian tumors in children and young adults. Arch Gynecol Obstet 2012;285(2):469–71. https://doi.org/10.1007/s00404-011-1946-2.

[15] Schorge JO, Eisenhauer EE, Chi DS. Current surgical management of ovarian cancer. Hematol Oncol Clin N Am 2012;26(1):93–109. https://doi.org/10.1016/j.hoc.2011.10.004.

[16] Zhao D, Song Y, Zhang Y, Li B. Outcomes of fertility-sparing surgery in ovarian juvenile granulosa cell tumor. Int J Gynecol Cancer 2019;29(4):787–91. https://doi.org/10.1136/ijgc-2018-000083.

[17] Oltmann SC, Garcia N, Barber R, Huang R, Hicks B, Fischer A. Can we preoperatively risk stratify ovarian masses for malignancy? J Pediatr Surg 2010;45(1):130–4. https://doi.org/10.1016/j.jpedsurg.2009.10.022.

[18] Fleming NA, de Nanassy J, Lawrence S, Black AY. Juvenile granulosa and theca cell tumor of the ovary as a rare cause of precocious puberty: case report and review of literature. J Pediatr Adolesc Gynecol 2010;23(4):e127–31. https://doi.org/10.1016/j.jpag.2010.01.003.