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A RARE CAUSE OF GROSS ASCITES IN AN EIGHT MONTHS OLD CHILD: JUVENILE GRANULOSA CELL TUMOR OF OVARY WITH ABNORMAL IMMUNOHISTOCHEMISTRY.

Histopathology

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ABSTRACT

Backgound: Granulosa cell tumors are rare, hormone secreting sex cord-stromal tumors accounting for 1% to 2% of all ovarian malignancies which exits in adult (95%) and juvenile (5%) forms. Presentation as gross ascites in an infant is further rare.

Case report: We present an eight months old child presenting as abdominal distension and diarrhea. Laboratory findings showed elevated levels of alfa-fetoprotein (AFP) only. Radiology revealed a solid-cystic ovarian mass in left ovary for which salphingo-ophrectomy was done. On histopathology a diagnosis of juvenile granulosa cell tumor which was negative for inhibin and calretinin.

Summary and conclusion: JCGT can present with elevated levels of AFP which can betray its stromal origin and create confusion with germ cell tumors clinically. Also, ovarian malignancy should be kept as a differential for ascites in infants.

KEYWORDS

Juvenile granulose cell tumor, infant, ascites, alfa- fetoprotein.

Background:

Granulosa cell tumors of ovary were first described by Rokitansky in 1855.[1] They are rare benign sex cord-stromal tumors (SCST) with an overall incidence of 0.4 to 1.7 cases per 100,000 women.[2] Among these juvenile granulosa cell tumor (JGCT) is even rarer and it presents before 30 years of age. As JGCT is a hormone producing tumor, it usually presents with symptoms like vaginal bleeding, pseudo puberty, irregular menstruation due to estrogen production and rarely hirsutism or virilization due to androgen production. Also, abdominal pain and abdominal distention can be present clinically. Ascites as a clinical manifestation is reported only in two cases previously.[3,4]

Case report:

An eight months old child presented with a history of abdominal distension and diarrhea for last one week. On clinical examination, her abdomen was hugely distended and tense with diffuse tenderness was present. However, no lump/mass was palpable separately. An abdominopelvic ultrasound was done which revealed gross ascites, along with a large solid cystic abdominopelvic mass measuring 10.7x8.5x9 cm with hypoechoic and hyperechoic foci within with increased internal vascularity in the left ovary. The left ovary was not visible separately. A probable diagnosis of germ cell tumor was reported by radiology. The serum level of alfa-fetoprotein (AFP) was mildly elevated (APF- 29.20 ng/ml, normal range-20 ng/ml). Ascitic fluid was sent for cytologic analysis, which showed no atypical cells and was negative for malignant cells. An exploratory laprotomy and tumor excision was planned. Left salpingo-oophorectomy was done. Grossly, the left ovarian mass was measured 10x8x8 cm. On gross examination, the mass was solid cystic filled with clear fluid (Figure 1). On microscopy, the round to oval tumor cells were arranged in sheets and follicle filled with eosinophilic material. The tumor cells had abundant eosinophilic cytoplasm. Frequent mitosis were noted. No nuclear grooves or schiller duval bodies were seen (Figure 2). On immunohistochemistry, the tumor cells were positive for CD99 and negative for AFP, PLAP, CD30, inhibin and calretinin (Figure 3). The morphology and immunohistochemistry were suggestive of juvenile granulosa cell tumor of left ovary, stage IA. Adjuvant therapy cisplatin was given for five days for 3 cycles one month apart. She had vaginal bleeding for 2 days post operatively which was self resolved. On follow-up the child is doing well and symptom free for last five months.

DISCUSSION:

Ovarian malignancies are very rare in infants. Granulosa cell tumor exits in two forms: Adult and juvenile. This classification is not only based on age of presentation, but also on the histolological features and natural history. Age is not the only criteria to distinguish these two since either type can occur at any age.[5] The mean age of presentation is 13 years but the present case was very young that is only eight months. Less than 20 cases of JGCT in infants are reported in the literature. Most of the cases of JGCT present with features related to hormone activity, such as irregular vaginal bleeding, puberty changes, somatoskeletal changes, hirisutism and virilism.[6]However, no such features were seen in our case. Our case presented solely with ascites. To the best of our knowledge, only two cases presenting only with ascites have been described in literature by Dhaiban et al. and Ashanagar et al.

The typical tumor size reported in the literature is more than 10 cms but it can vary from 3 to 25 cms. Tumor markers like inhibin can be elevated in JGCT, but in our case AFP was done and found to be elevated due to clinicoradiological suspicion of germ cell tumor. AFP is a tumor marker of Germ cell tumors, but it has been found to be raised in some of the non germ cell tumors like epithelial tumors of ovary and sex cord tumors, mainly sertoli- leydig cell tumors. On literature review we found that juvenile granulosa cells tumors can also present with increased levels of AFP.[7] Of note, the tumor cells were negative for AFP using immunohistochemistry. The reason for this remains unexplained. Possibly, this raised AFP can be due to ascitis because AFP was not positive on immunohistochemistry and it was only slightly raised.[8] Also, the tumor cells were negative for both calretinin and inhibin. The morphology was consistent with juvenile granulosa cell tumor only and no other germ cell component was identified even on extensive sectioning.

The tumor was staged using the International Federation of Gynecology and Obstetrics (FIGO) system. Surgery is the mainstay of the treatment. Adjuvant chemotherapy or observation (category 2B recommendation as per NCCN) is indicated for stage Ic, IIIc or for tumors with high mitotic rate. In the present case, cisplatin was given because of high mitotic count of the tumor. The recommended combination chemotherapy was not given due to young age of the patient.

JGCT has a very good prognosis. Five year survival rate is 90-95% for stage I tumors and 20 to 25 % for advanced tumors. The good prognostic factors include: less than 10 years of age, presence of precocious pseudopuberty, FIGO stage I and FOXL2 expression, whereas the bad prognostic factors include: extra-capsular extension of tumor within ovary, tumor rupture, nuclear atypia, high mitotic rate and presence of residual disease after surgery. Recurrence, although rare, can occur in first 48 months post diagnosis and in a very few cases even after many years.[9]These patients require a long term follow up.

CONCLUSION:

Although ovarian tumors are very rare in infants they should be kept in the differential of gross ascitis. Radiology and tumor markers are quite useful for differentiating germ cell tumors from other ovarian tumors, but they can clinically create a diagnostic dilemma as in the present case. Histopathology remains the gold standard for definitive diagnosis.

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Conflicts of interest: None.

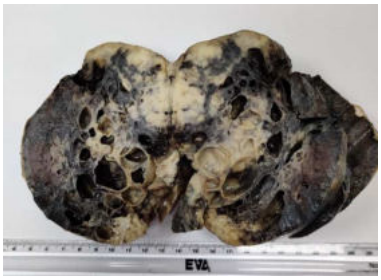

Figure 1. Gross photograph showing solid cystic grey brown tumor in left ovary.(post formalin fixation)

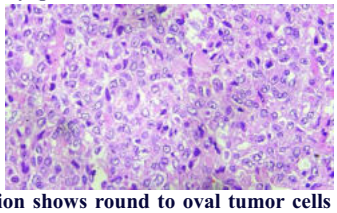

Figure. 2. Section shows round to oval tumor cells in nests and follicles with eosinophillic cytoplasm. (H& E stain, 400X)

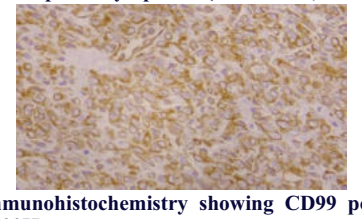

Figure. 3. Immunohistochemistry showing CD99 positivity in tumor cells. (400X)

REFERENCES:

1. Rokitansky CV: Über Abnormalitäten des Corpus Luteum. Wien Med Ztc, 1859;4:253-4.
2. Calcaterra V, Nakib G, Pelizzo G et al: Central precocious puberty and granulosa cell ovarian tumor in an 8-year old female. Pediatr Rep, 2013; 5(3): e13
3. Dhaiban M, Akhtar N, Asif M, Tanwani A. Ascites as a Sole Presentation of Juvenile Granulosa Cell Tumor. J Pediatr Neonatal Care, 2015; 7 3(2): 00113.
4. Ashnagar A, Alavi S, Nilipour Y, Azma R,Falahati F. Massive Ascites as the Only Sign of Ovarian Juvenile Granulosa Cell Tumor in an Adolescent: A Case Report and a Review of the Literature. Case Reports In Oncology Medicine, 2013; 1:1-4.
5. Biscotti CV, Kennedy AW. Ovarian Juvenile Granulosa Cell Tumors**.** Adolesc Pediatr Gynecol .1990;3:15-19.
6. Bus D, Buzogány M, Nagy G, et al. Rare virilizing granulosa cell tumor in an adolescent. Mol Clin Oncol 2017;6:88–90.
7. Smith R, Moss J, Shore I, El-Bahrawy MA. Juvenile granulosa cell tumour with hepatocyte-like cells and raised serum alpha-fetoprotein. Histopathology 2010;57:637-41
8. Frausto SD, Geisler JP, Fletcher MS, et al: Late recurrence of juvenile granulosa cell tumor of the ovary. Am J Obstet Gynecol 2004; 191:366e7
9. Zhu F-L, Ling A-S, Wei Q, Ma J, Lu G. Tumor Markers in Serum and Ascites in the Diagnosis of Benign and Malignant Ascites. Asian Pacific Journal of Cancer Prevention [Internet]. 2015 Feb 25;16(2):719–22.