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JUVENILE GRANULOSA CELL TUMOR A NEO-NATAL REVELATION

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ABSTRACT

The juvenile granulosa cell tumors of the ovary belong to the group of stroma and sex cord tumors. They are rare, representing 1 - 2% of ovarian tumors. They are involved in ovarian hormone function, responsible for the synthesis of hormones (estrogen, corticosteroids), which explains the hormonal dysfunctions associated with these cancers. The juvenile granulosa cell tumors occur in older children, usually before the age of 20 years old, with a maximum frequency in pre-puberty period. We report an exceptional case of a granulosa tumor in a four-month infant.

Keywords: Juvenile Granulosa cell tumors, Immunohistochemistry, CT scan, Neo-natal.

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OBSERVATION

A 4- month old infant was admitted at the emergency department for abdominal bloating. In the medical history, the pregnancy was not attended, with a vaginal delivery without anomalies. A physical examination found an infant in a good condition with a distended abdomen and diffuse hyperpilosity predominating at face, pubisand the limbs.

Abdominal ultrasonography (**Figure1**) revealed ascitis with a right large, solid and heterogeneous ovarian mass measuring 43 x 35 mm, a pubescentlike uterus measuring 44 x 14 x 28 mm with clearly individualized line of emptiness and a ratio body / cervix>1. The left ovary size had increased. The liver, spleen, kidneys were without any abnormalities.

The abdomino-pelvic CT scan (**Figure 2**) showed a right latero-uterine mass, of tissue density, heterogeneously elevated with abundant ascitis. It did not reveal any deep lymph nodes.

Hormonal Dosages revealed increased levels of estrogens with normal alpha fetoprotein and beta chorionic gonadotrophin hormone.

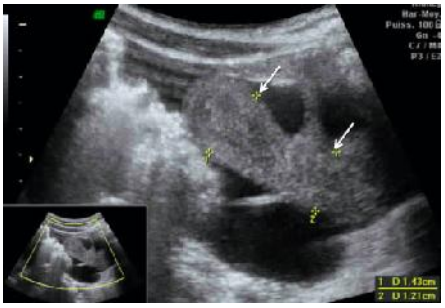

Figure 1: 4 months infant. Ultrasonography findings: Pubertal type uterus with emptiness line clearly individualized and thick endometrium. Note the presence of ascitis +++.

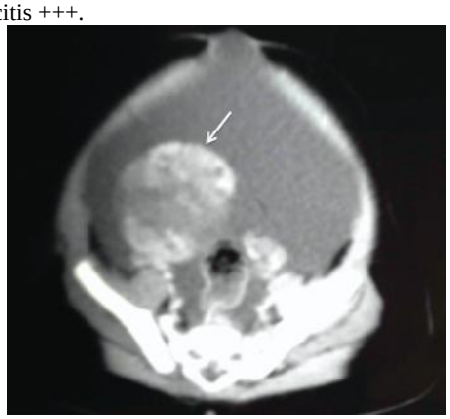

Figure 2: 4 months infant. CT scan (axial reconstruction): Abundant ascitis with right heterogeneous ovarian mass without calcification or fat areas; Puberty type homogeneous uterus. Left ovary is normal. Absence of deep nodes.

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The diagnosis of the right secreting ovarian tumor was discussed and the possibility of a granulosa cell tumor was raised. The therapeutic decision adopted was elective surgery which consisted to a right oophorectomy associated with a partial amputation of the homolateral fallopian tube. The histopathologic examination of the surgical specimen (**Figure 3 and 4**) coupled to the immunohistochemical study confirmed the diagnosis of juvenile granulosa cell tumor with a luteinization phenomenon. The postsurgical course was uneventful. At the follow-up, regular ultrasound examination (**Figure 5**) and biological controls over a period of six months were normal.

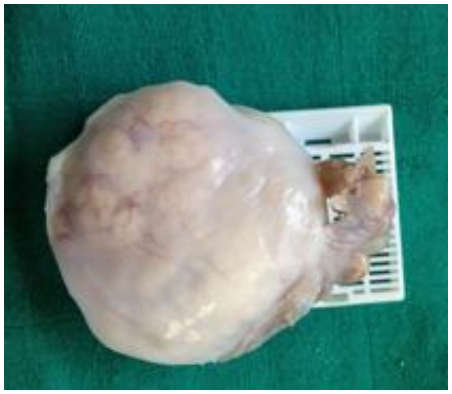

Figure 3: Macroscopic appearance of surgical specimen of right oophorectomy; closed and open shows a yellowish solid mass firm with grayish white zones and a cystic formation with serous contents.

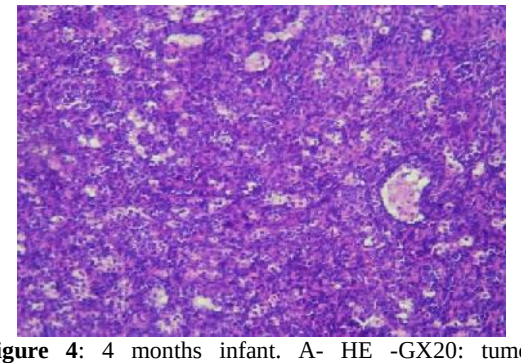

Figure 4: 4 months infant. A- HE -GX20: tumor proliferation of round or polygonal cells showing a pseudo follicular and a luteinization phenomenon (clarified quota cell cytoplasm). B- HE -GX40: Pseudo follicular appearance with the presence of macrophages under the light.

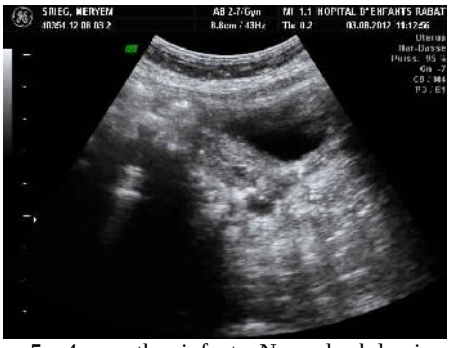

Figure 5: 4 months infant. Normal abdominopelvic ultrasound scan checkup.

DISCUSSION

The granulosa cell tumors are rare, representing 1-2% of all ovarian tumors [1]. They are often secreting functional lesions. There are two distinct clinicopathological entities: the adult granulosa cell tumor, the most common up to 95% of cases and juvenile granulosa cell tumor representing less than 5% [2, 3]. The juvenile form usually occurs before the age of 20 [3], with a maximum frequency in pre puberty period [1, 4]. The discovery of this type of tumor in the neonatal period has never been described in literature. To our knowledge this is the first reported case. The early onset of the granulosa cell tumor in our patient leads us to ask about the possibility of a pre natal development of this tumor.
Clinically, granulosa cell tumors occur with a tumor syndrome: abdominal distension with ascitis of varying importance depending on the size of the tumor. Sometimes it is an endocrine syndrome associated with excretory functions of these tumors which is indicative: early pseudo puberty [3, 6].
Imaging of granulosa cell tumors is polymorphic, non-specific: With the ultrasound scan, the juvenile granulosa cell tumor appears generally as an important ovarian mass, with mixed solid and cystic content, sometimes with a multilocular aspect. The cystic walls may have an echogenic content related to the occurrence of bleeding. The Doppler, when performed, reveals an abundant vascularisation of the walls and in the tissular part. The presence of an associated endometrial thickening is an important guidance for the diagnosis of granulosa cell tumor [2,7]. It shows the presence of hyperestrogeny. In computerized tomography (CT), the lesion is tissual or mixed, usually with no calcifications or fat component. PC injection shows an early enhancing of the fleshy part and the septa.
In magnetic resonance imaging (MRI) the appearance is highly polymorphic, sometimes the mass is hard, sometimes cystic, multilocular, more often mixed. The T1 hyperintense is of granulose

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tumor is often related to the hemorrhagic component. The dynamic MRI injected shows a type 2 or 3 curve [4, 8]. Concerning biological parameters, the estradiol dosed in case of early puberty is high. The inhibin is currently the specific marker of granulosa cell tumors.

The pathologic analysis confirmed the diagnosis of granulosa cell tumors showing a proliferation of round or polygonal cells with little eosinophilic cytoplasm and granular chromatin nuclei without atypia. These cells are organized into tracks with pseudo follicular cavities. The stroma is readily luteinized. The immune-histochemical study shows an expression of inhibin-alpha which has a diagnostic value.

The first-line treatment of granulosa cell tumors is surgical: adnexectomy in the juvenile form of IA FIGO stage, total hysterectomy in adult granulosa cell tumors [2, 3]. Chemotherapy is available in case of IC stage or stage II tumor, and in case of recidivism further to reoperation [1, 9, 10].

CONCLUSION

The Juvenile Granulosa cell tumor is a very rare entity. It usually occurs before the age of 20, with a maximum frequency in pre-puberty period. A neonatal diagnosis is unusual. Imaging can suggest the diagnosis in case of a cystic or mixed ovarian mass associated with endometrial hyperplasia. Positive diagnosis is histological.

There is no conflict of interest.

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