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Case Report

Ovarian Juvenile Granulosa Cell Tumor in Childhood: The Importance of Early Diagnosis. Case Report

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Abstract

Introduction: Juvenile granulosa cell tumors (JGCT) of ovary are very rare among children. They are typically seen within the first two decades of life with signs of hyperestrogenism and abdominal mass. In childhood, the diagnosis should be suspected in girl with symptoms of pubertal development.

Objective: We report a rare case of a 2.5 year-old girl with peripheral precocious puberty due to a JGCT of left ovary and we highlight the keys to diagnosis and treatment of this tumor.

Clinical Case: 2.5 year-old female child with developing breasts, pubic hair and vaginal bleeding along with a palpable abdominal mass presented to our pediatric surgery department.

Ultrasound showed a left adnexal lesion that contained both solid and cystic components. Doppler ultrasound study revealed vascularity within the lesion. MRI showed a large cystic and solid abdominopelvic mass measuring 12 cm that reached the subrenal aorta, probably originating from the left ovary.

Serum levels of βHCG, αFP, CA19-9 and CA125 were normal. In view of clinical diagnosis of ovarian tumor, a chest X-ray and a thoracoabdominal CT scan were also performed and showed no secondary tumors.

Exploratory laparotomy was then done through a Pfannenstiel incision with evidence in the pelvis of a 15 cm mass of the left ovary that weighed about 650 grams and had a very rich vascularization. Contralateral ovary and uterus were normal. A left adnexectomy was carried out and tumour was sent for histopathological examination. The biopsy showed TCGJ of left ovary with FIGO Stage IA. Immunohistochemical study reported a diffuse immunoreactivity for Inhibin.

There was no evidence of recurrence within an 18-month follow-up, however, careful observation may be required as there is a possibility of tumour recurrence.

Conclusion: Juvenile granulosa cell tumors are very uncommon among the pediatric population. Most of the TCGJ are diagnosed in stage I. In early stages of the tumor, surgery is curative and the prognosis on a long-term basis is favourable, but a strict surveillance of images and tumoral markers is needed. The most specific marker is Inhibin B. Chemotherapy may be required in advanced stages.

Keywords: *Juvenile Granulosa Cell Ovarian Tumor; Peripheral Precocious Puberty; Inhibin B*

Introduction

Juvenile granulosa cell tumor (JGCT) is a rare tumor in infants. The diagnosis must be considered in all ovarian tumors of children, especially when they present with early signs of puberty. This neoplasm has favorable prognosis if diagnosed at an early stage. Presenting symptoms of this tumor are diverse and the definitive diagnosis relies on histopathology.

Case Study

A 2.5 year-old girl, with no medical history, presented to our department with abdominal distension and early signs of puberty since 2 months: developing breasts, pubic hair and vaginal bleeding.

Physical examination showed brown freckling spots on the right thigh and a firm and mobile abdominal mass measuring 9 cm. Puberty signs were between Tanner stages 2 and 3.

Abdominal ultrasonography revealed a left adnexal lesion that contained both solid and cystic components. Doppler ultrasound study revealed vascularity within the lesion.

MRI showed a large cystic and solid abdominopelvic mass measuring 12 cm that reached the subrenal aorta, probably originating from the left ovary.

Serum levels of βHCG, αFP, CA19-9 and CA125 were normal.

In view of clinical diagnosis of ovarian tumor, a chest X-ray and a thoracoabdominal CT scan were performed and showed no secondary tumors. Exploratory laparotomy was then done through a Pfannenstiel incision with evidence in the pelvis of a 15-cm mass of the left ovary that weighed about 650 grams and had a very rich vascularization. A left adnexectomy was performed and the specimen was sent for histopathology.

Anatomopathological study revealed a JGCT of left ovary with FIGO Stage IA. Immunohistochemical study reported a diffuse immunoreactivity for Inhibin.

The post-operative course was uneventful with an 18-month follow-up.

Discussion

Granulosa cell tumors (GCTs) are rare sex cord–stromal tumors accounting for 1% to 5% of all ovarian tumors. GCT of ovary was first described in 1985 by Rokitansky. Juvenile GCT differs from the adult [1]. Almost 80% of JGCTs occur before age 20 [2] and the mean age at diagnosis is 13 years. In our case, the patient was a 2.5 year-old girl.

The most common symptoms are abdominal pain and distension. In our case, the main symptoms were abdominal distension and signs of precocious puberty.

Generally, JGCT is diagnosed in stage I as in our case, according to the FIGO staging for ovarian cancer. Stage I corresponds to a tumor limited to the ovaries. The mortality in this stage is very low and patients do not require complementary therapies and have a good prognosis on a 5-year term (90 - 100%) [3,4].

The clinical and pathological features of JGCTs are diverse, and there is no standard treatment. However, primary management of this tumor is surgery and the benefit of adjuvant chemotherapy is unclear. Unilateral salpingo-oophorectomy is the treatment for children and women in the reproductive age with stage IA disease.

In females, ovarian granulosa cells produce Inhibin B, a member of TGFβ family of growth factors. It is thought that Inhibin B is a reliable marker to detect recurrence during the follow-up period [5].

The prognosis of infancy patients seems to be better than that of adult patients because infancy patients often develop hormone-related symptoms and are diagnosed at an early stage [6].

Conclusion

JGCT is a rare neoplasm with different clinical behavior and is easily confused with other tumors. The primary management is through surgery. JGCT has a favorable prognosis in patients with stage I disease managed by surgery. Adjuvant chemotherapy may be indicated in advanced stages. A follow-up with Inhibin B is recommended.

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