


Ovarian Juvenile Granulosa Cell Tumor in A 2.5 Years-Old Girl: The Importance of Early Diagnosis

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Abstract

Juvenile granulosa tumor is a rare tumor in infant. The diagnosis should be discussed in any ovarian tumor of children, especially in front of the early signs of puberty. The prognosis of granulosa cell tumors remains good. The main prognostic factor is the stage of local extension. Early diagnosis at early signs of puberty is an important prognostic factor. We describe the case of a girl aged 2 years and 5 months who presented signs of early puberty. The diagnosis was a secreting ovarian tumor but was very large at that time. We focus by this observation on the value of early diagnosis of this tumor type which remains a key prognostic factor.

Keywords

- Ovary
- juvenile granulosa tumor
- Early puberty
- Infant

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 Report:

A 2.5 years-old girl, with no medical history, presented to our department with abdominal distension and early signs of puberty for 2 months: developing breasts, pubic hair and vaginal bleeding (**Figure 1**).



Figure 1: Abdominal distension and pubic hair

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

Abdominal ultrasonography (USG) revealed a left adnexal lesion that contained both solid and cystic components. Doppler USG 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 β HCG, α FP, 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 done through a Pfannenstiel incision with evidence of a 15-cm mass of the left ovary that weighed about 650 grams and had a very rich vascularization (**Figure 2-3**). A left adnexectomy was performed and the specimen was sent for histopathology.

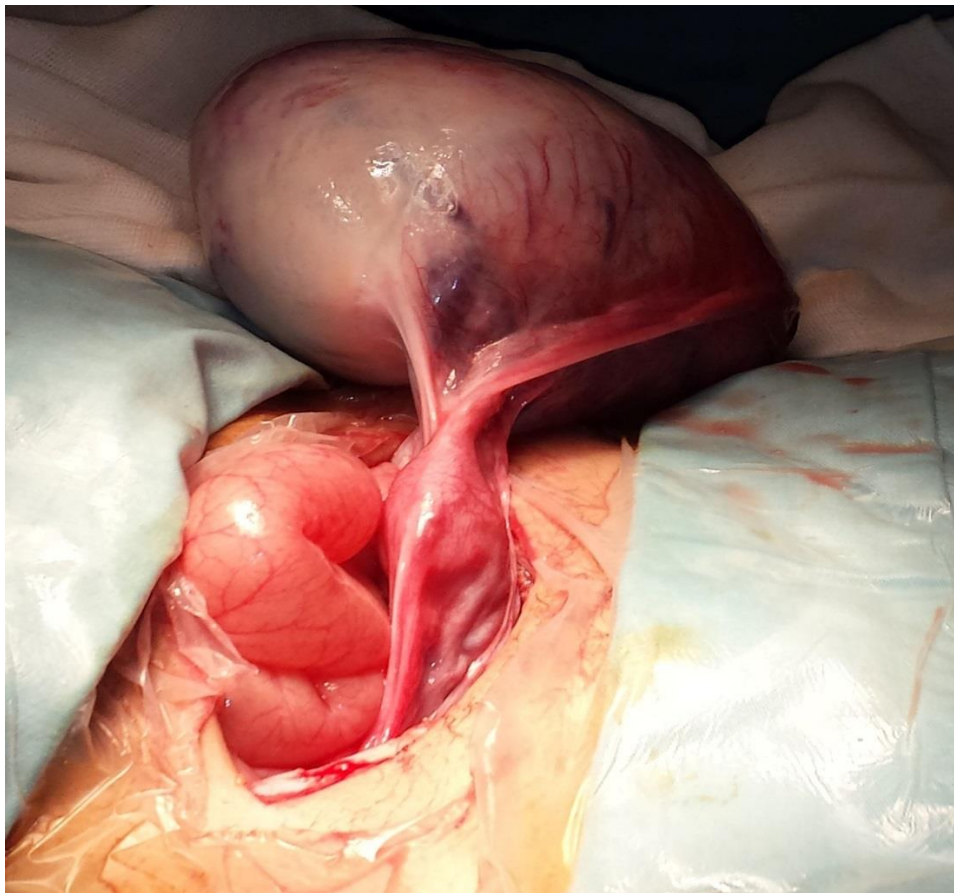


Figure 2: Ovarian tumor



Figure 3: Adnexectomy

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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 20² and the mean age at diagnosis is 13 years. In our case, the patient was a 2.5 years-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 IA as in our case. 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, a member of TGF β family of growth factors. It is thought that inhibin B is a reliable marker to detect recurrence during the follow-up period.³

The prognosis of infants seems to be better than that of adult patients because infants often develop hormone-related symptoms and are diagnosed at an early stage.⁴

JGCT is a rare neoplasm with different clinical behavior and is easily confused with other tumors. The primary management is the surgery. JGCT has a favorable prognosis in patients with stage I disease managed by surgery. Adjuvant chemotherapy may be indicated in higher stages.

Conclusion:

We focus by this observation on the value of early diagnosis of this tumor type which remains a key prognostic factor.

Ethical Consideration

This study was approved by Habib thameur Hospital, pediatric surgery department, Tunis, Tunisia.

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Conflict of interests

There is no conflict of interest

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