

CAS CLINIQUE / CASE REPORT

PRECOCIOUS PUBERTY SECONDARY TO AN OVARIAN CYST

A Case Report

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ABSTRACT • Introduction : The functional ovarian cyst remains a rare disease in the prepubertal population; it is however a leading cause of precocious pseudo-puberty. Precocious puberty results in significant changes marked by growth acceleration and premature closure of growth plates. We report in this article the case of a precocious pseudo-puberty secondary to a functional ovarian cyst.

Case presentation : A six and a half years old patient, consulted for a recent appearance of early breast development. Hormonal analysis revealed a distal hormonal secretion with suppression of the hypothalamic-pituitary axis. Pelvic ultrasound revealed the presence of a large right ovarian cyst, confirmed by pelvic MRI. The management of this patient consisted in a simple monitoring. After two months, a complete regression of the symptoms was noted as well as a complete disappearance of the ovarian cyst on ultrasound, and an elimination of the hypothalamic-pituitary axis suppression on hormonal analysis.

Conclusion : Ovarian cysts are mostly self-limited and require no treatment with spontaneous regression. In rare cases surgery is necessary; it is important to consider a conservative intervention of ovarian tissue without affecting fertility. Ovarian cysts are rare in the pediatric population and with good prognosis in their majority; they require however the attention of the pediatrician, given the possible risk of malignancy.

Keywords : ovarian tumors, precocious puberty, functional cysts

INTRODUCTION

Precocious puberty is defined by the development of secondary sexual characteristics before the age of eight years in girls. This results in significant changes marked by growth acceleration and premature closure of growth plates.

In most cases, early puberty is of central origin, rarely, there is a peripheral hormonal secretion or pseudo-precocious puberty.

The functional ovarian cyst remains a rare disease in the prepubertal population, however, it represents the

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RÉSUMÉ • Le kyste fonctionnel de l'ovaire reste une maladie rare dans la population prépubère, il est cependant une des causes majeures de pseudo-puberté précoce. La puberté précoce entraîne des modifications importantes marquées par l'accélération de la croissance et la fermeture prématurée des cartilages de croissance. Nous rapportons ici un cas de pseudo-puberté précoce secondaire à un kyste ovarien fonctionnel.

Présentation du cas : Patiente âgée de six ans et demi, consultant pour l'apparition récente de signes de puberté. Le bilan hormonal révèle une sécrétion hormonale distale avec suppression de l'axe hypothalamo-hypophysaire. L'échographie pelvienne révèle la présence d'un kyste de l'ovaire droit, confirmée par IRM pelvienne. La prise en charge a consisté en une simple surveillance. Après deux mois, une régression complète des symptômes a été notée, ainsi qu'une disparition totale du kyste ovarien à l'échographie et une élimination de la suppression de l'axe hypothalamo-hypophysaire sur le bilan hormonal.

Conclusion: Les kystes ovariens sont souvent auto-limités avec régression spontanée, et ne nécessitent aucun traitement. Dans de rares cas, la chirurgie est nécessaire, et il est important d'envisager une intervention conservatrice du tissu ovarien sans affecter la fertilité. Les kystes ovariens sont rares dans la population pédiatrique et de bon pronostic dans leur majorité, mais ils exigent l'attention du pédiatre, étant donné l'impact possible du retard diagnostique sur le pronostic, la croissance et la fertilité.

Mots-clés : tumeurs ovariennes, puberté précoce, kystes fonctionnels

leading cause of pseudo-precocious puberty [1-2].

Through this article, we report the case of a precocious pseudo-puberty diagnosed in a six and a half year old girl that turns to be secondary to a functional ovarian cyst.

CASE PRESENTATION

We present a six and a half years old patient without any past medical history and with normal height and weight development, consulting for a recent appearance of early signs of puberty, including a breast development and vaginal secretions, without acceleration of growth velocity.

Physical examination showed:

- Height 117.5 cm (+0.5 SD); weight 18 Kg (-1 SD)
- Nipples raised indurate and painful, with absence of axillary and pubic hair (A1P1S2)
- Nothing relevant for the rest of the physical exam

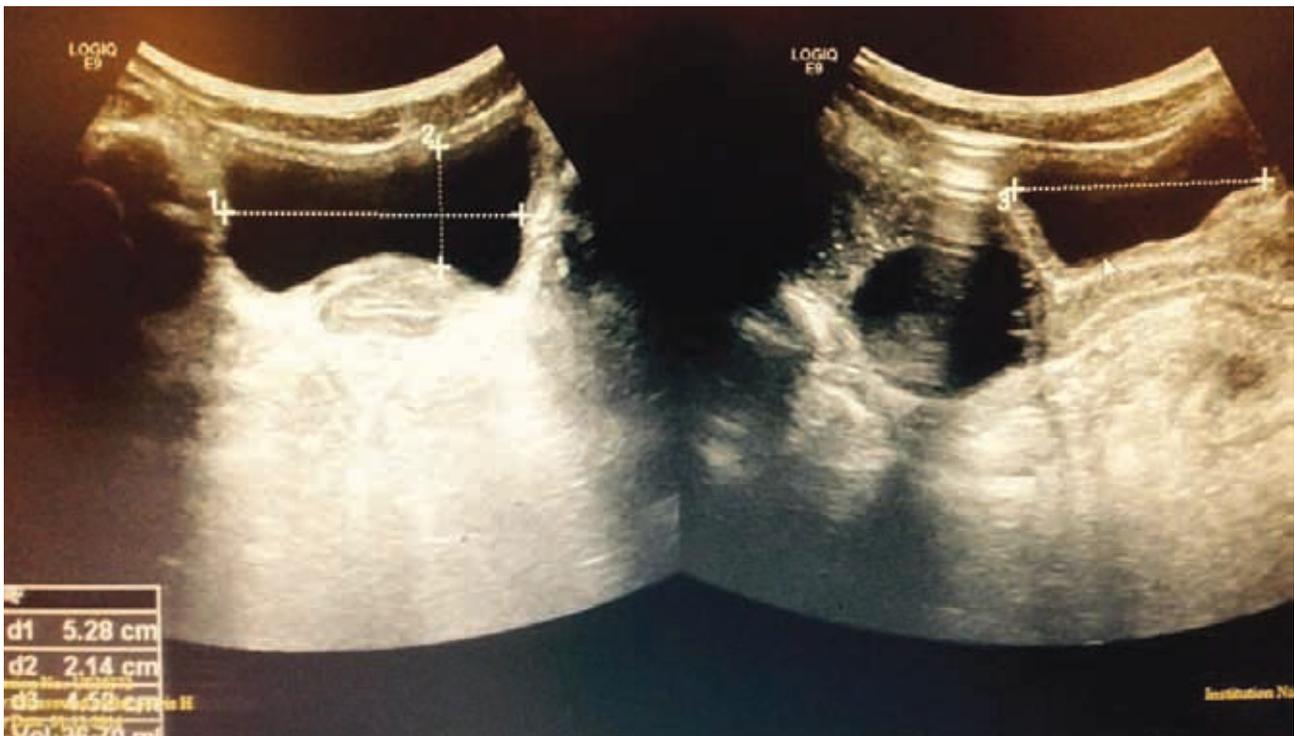


Figure 1. Right ovarian cyst on pelvic ultrasound

especially neurological exam, no “café au lait” spots and no history or actual bone fractures.

Regarding the suspicion of precocious puberty, an initial assessment was done:

- A bone age estimated by Greulich and Pyle atlas, turned to be seven and a half;
- Hormonal analysis* revealed a distal estradiol secretion with suppression of the hypothalamic-pituitary axis: FSH 0.14 pg/ml; LH < 0.1 pg/ml; estradiol 566 pg/ml;
- The GnRH stimulation test was not performed because of its non availability in our center.

The assessment is then completed for etiologic diagnosis:

- Pelvic ultrasound revealed the presence of a 36 x 34 x 27 mm right ovarian cyst. (Figure 1)
- Pelvic MRI, realized for a more accurate assessment of the cyst, revealed a thin endometrium measuring 2 mm, a 2.7 x 1.2 x 0.8 cm left ovary containing few follicles, and the presence of a large cystic structure occupying the right ovary measuring 4.2 x 2.5 x 3.8 cm with minimal peripheral enhancement after gadolinium administration. (Figure 2)

Delta 4 androstenedione and dehydroepiandrosterone (DHEA) dosage showed no abnormalities, thereby eliminating a congenital adrenal hyperplasia.

In order to rule out malignancy cause of the cyst, the work-up was completed by:

- Beta HCG, CA 125 and alpha fetoprotein all showing normal levels for the age;
- Abdominal ultrasound: no abnormality.

* FSH: follicle-stimulating hormone LH: luteinizing hormone

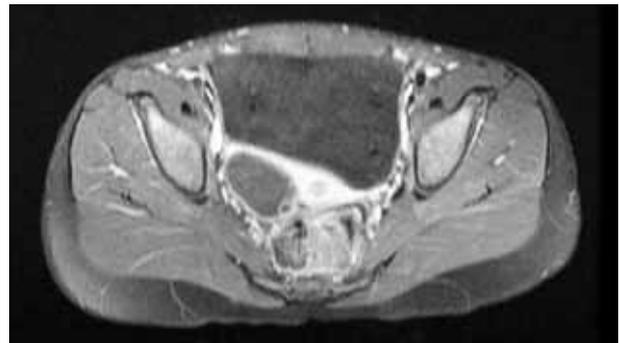


Figure 2. Right ovarian cyst on pelvic MRI

The management of this patient consisted in a simple monitoring.

After two months, a complete regression of the symptoms was noted as well as a complete disappearance of the ovarian cyst on ultrasound. Furthermore, hormonal analysis demonstrated a significant drop in the rate of estradiol that became undetectable (< 5 pg/ml) and an elimination of the hypothalamic-pituitary axis suppression with FSH level increasing to 2.01 pg/ml.

DISCUSSION

Even though ovarian cysts responsible of precocious puberty represent a well-known condition in the pediatric population, it is important to mention that the publication of this article is an opportunity to seek awareness of pediatricians on this subject, considering its rarity and the risk of associated malignancy.

Precocious puberty in girls is defined by the appearance of secondary sexual characteristics (breast development, growth spurt, appearance of pubic hair or in axillary) before the age of eight years [3]. There are two forms of hypergonadism: central hypergonadism dependant on gonadotropins, and peripheral hypergonadism or precocious pseudo-puberty, secondary to peripheral hormonal secretion or congenital adrenal hyperplasia.

The GnRH stimulation test is currently considered the gold standard test in the diagnosis of precocious puberty [4]. It can distinguish between early puberty of central origin as FSH and LH increase in response to this stimulation while their suppression persists in case of early pseudo-puberty [5].

A baseline level of gonadotrophin can be sufficient in case of very high levels of estradiol.

Peripheral hypergonadism is found in case of functional ovarian cysts or malignancies with secretory component.

Ovarian tumors are mostly observed in the adult population, their occurrence in children is relatively rare and most often, they consist of benign and functional cysts [6].

The simple ovarian cyst is the most common tumor in the pediatric population in all age groups. It consists, in most cases, of functional cysts which develop probably secondary to intermittent gonadotrophin secretion [7-8].

Clinical manifestations depend on the nature of the tumor, its size and its hormonal secretion. They include abdominal and pelvic pain that may be related to the pelvic mass, cyst rupture, the intracystic hemorrhage or the ovarian torsion. The abdominal mass palpation on physical examination is also possible in case of large tumor size.

In this regard, we note as an example the case report of a 14-year-old premenarchal girl presenting chronic abdominal pain, constipation and abdominal enlargement, that turned to be secondary to a 9 Kg ovarian mucinous cystadenoma removed surgically by laparotomy with salpingo-oophorectomy [9].

As part of functional ovarian cysts, it is important to mention the Mc Cune Albright syndrome defined by a clinical triad consisting, in addition to functional ovarian cysts, of fibrous bone dysplasia and "café au lait" cutaneous spots. Note that the development of ovarian cyst can precede other clinical manifestations of this syndrome as it can be recurrent. Thus, in case of clinical suspicion, a molecular DNA analysis must be indicated to confirm the diagnosis. Mc Cune Albright syndrome is caused by a mutation of the alpha subunit of the G protein encoded by the gene GNAS1 [10].

The thyroid stimulating hormone was not tested in the case of this patient due to the absence of signs of hypothyroidism; however, profound hypothyroidism of long duration can be associated with precocious puberty [11].

In addition, thyroid abnormalities can be associated with McCune Albright syndrome [12].

In general, ovarian cysts of less than 1 cm of diameter are asymptomatic, while early pseudo-puberty is secondary to cysts that have a diameter that exceeds 2 cm [1].

The case reported in this article shows an isolated increase in estradiol with suppression of the hypothalamic-pituitary axis, which is compatible with a pseudo-precocious puberty that turned to be secondary to an ovarian cyst.

The dosage of tumor markers is still required to rule out malignancies even though their prevalence remains minimal, estimated at less than 1% of all ovarian tumors, given the poor prognosis these tumors can present.

These tumor markers are:

- Beta HCG (associated with embryonal carcinoma and dysgerminoma);
- Alpha fetoprotein (associated with teratoma and tumors of yolk sac);
- CA125 (associated with epithelial tumors). [13]

Ovarian cysts are mostly self-limited and require no treatment with spontaneous regression after an average of 3 to 4 weeks [2].

In rare cases surgery is necessary; it is only indicated in the case of ovarian torsion or non regression of clinical symptoms after 3 months [14-15].

If surgery is indicated, it is important to consider a conservative intervention of ovarian tissue without affecting fertility.

In the case presented above, the treatment consisted in simple monitoring with spontaneous regression of clinical symptoms and radiological image after two months.

CONCLUSION

Although precocious puberty in girls is generally central and idiopathic, peripheral origin should remain in our minds. Functional cysts are the most frequent cause in this case, and have an excellent prognosis. Nevertheless, it is mandatory to eliminate a malignancy.

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