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Recurrent ovarian torsion in a girl with peripheral precocious puberty

Torsión ovárica recurrente en una niña con pubertad precoz periférica

Nuria López Barrena[1], Belén Sagastizabal Cardelús[1], María Luisa Lorente Jareño[2], Rubén Martín Alelu[3], Andrés José Alcaraz Romero[1]

[1]*Pediatría. Hospital Universitario de Getafe. Getafe, Madrid*
[2]*Radiología. Hospital Universitario de Getafe. Getafe, Madrid*
[3]*Cirugía Pediátrica. Hospital Universitario 12 de Octubre. Madrid*

Abstract

We present an exceptional case of a girl with peripheral precocious puberty probably due to an autonomous ovarian cyst and recurrent episodes of ovarian torsion in a 7-year-old girl who presented with thelarche and advanced bone age. In the clinical evaluation, she presented symptoms compatible with precocious puberty. However, hormonal studies were not conclusive of peripheral precocious puberty since we did not find an increase in sex steroids (FSH 0.7 mU/mL, LH <0.3 mU/mL, estradiol <5 pg/mL). She presented intermittent episodes of vaginal bleeding and abdominal pain. Abdominal imaging revealed a right oedematous ovary with multiple cysts in its periphery and an image that was compatible with a thickened angulated vascular pedicle (gonadal vein) indicative of torsion. She was treated with laparoscopic right ovarian detorsion and oophoropexy. A month and a half later, she presented a new episode of ovarian torsion and therefore had to undergo another operation.

Key Words: Recurrent ovarian torsion, Ovarian cysts, Precocious puberty, Oophoropexy

Resumen

Presentamos un caso excepcional de una niña con pubertad precoz periférica probablemente secundario a quistes ováricos autónomos y episodios recurrentes de torsión ovárica. Se trata de una niña de 7 años que nos remitieron a la consulta de endocrinologia pediátrica por telarquia y edad ósea avanzada. En la evaluación clínica presentó signos compatibles con pubertad precoz. Sin embargo, los estudios hormonales no fueron concluyentes de pubertad precoz periférica, ya que no encontramos aumento de esteroides sexuales (FSH: 0,7 mU/mL, LH < 0,3 mU/mL, estradiol < 5 pg/mL). Durante su seguimiento presentó episodios de sangrado vaginal intermitente y dolor abdominal. La prueba de imagen abdominal reveló un ovario edematoso derecho con múltiples quistes en su periferia y una imagen que se correspondía con un pedículo vascular (vena gonadal) angulado y engrosado, indicativo de torsión. Fue intervenida quirúrgicamente mediante detorsión ovárica derecha vía laparoscópica y ooforectmía. Un mes y medio más tarde presentó un nuevo episodio de torsión ovárica, por lo que tuvo que ser intervenida de nuevo.

Palabras clave: Torsión ovárica recurrente, Quistes ováricos, Pubertad precoz, Ooforectomía

Correspondencia:
Nuria López Barrena
Pediatría
Hospital Universitario de Getafe
Getafe, Madrid
E-mail: nlbarrena@hotmail.com

Introduction

Peripheral precocious puberty is defined as the appearance of secondary sexual characteristics not dependent on the hypothalamic-pituitary-gonadal

axis [1]. Its most common cause is ovarian cysts [2]. On the other hand, ovarian cysts have been associated with an increased risk of ovarian torsion. Ovarian torsion is a pathology considered a surgical emergency [3]. Early diagnosis and treatment are required to avoid loss of adnexa and other complications. After the intervention, recurrence is rare and management options include oophoropexy after detorsion [4]. We describe an exceptional case of a girl with precocious puberty who suffered from intermittent vaginal bleeding and recurrent episodes of ovarian torsion.

Case Description

A girl aged 6 years and 8 months was referred to the paediatric endocrinology clinic for left unilateral thelarche. Advanced bone age: 8 years and 5 months old (Greulich & Pyle). Abdomino-pelvic ultrasound: both ovaries enlarged (right ovary 13 cc, left 15 cc) with follicular cysts (<9 mm). At 7 years and 5 months, a pubertal development in TANNER stage III (S3, P1, A1) was observed with a prepubertal response to the requested GnRH stimulation test. No cafe au lait patches on the skin or cystic changes in bones were found. At 8 years and 2 months old, she presented self-limited vaginal bleeding with a new episode at 8 years and 6 months old, this time accompanied by abdominal pain and vomiting. A new abdominal ultrasound was requested, showing an enlarged right ovary with a volume of 185.5 cc (10 x 5 x 6 cm) with a solid, rounded appearance and multiple cysts of up to 15 mm, as well as a uterus with an infantile morphology and left ovary 27.22 cc (2.5 x 4.5 x 3 cm), oval with follicular cysts (<10 mm) (Figure 1). Abdominal magnetic resonance imaging was performed confirming a right ovarian tumour with an image consistent with thickened angulated vascular pedicle (gonadal vein), indicative of torsion. She presented an oedematous ovary with multiple cysts in its periphery (the largest measuring 15 mm) and free fluid in the pelvis (Figure 2). Blood test: FSH 0.7 mU/mL, LH <0.3 mU/mL, estradiol <5 pg/mL. Tumour markers were negative. The operation was performed laparoscopically. During the operation, a very enlarged right ovary, with pedicle torsion, was observed. Throughout the procedure, an improvement in the colouration of the adnexa was observed. The operation involved detorsion of the ovary (2 turns) and right oophoropexy. No cystectomy was performed. Histopathological examination of peritoneal fluid was negative for malignant cells and showed acute and chronic inflammation and reactive mesothelial hyperplasia. A control ultrasound was performed 15 days post-intervention with ovarian reduction (60 cc). A month and a half later, she presented a new episode of abdominal pain and vomiting. Abdominal-pelvic ultrasound was repeated, showing a new increase in right ovarian volume

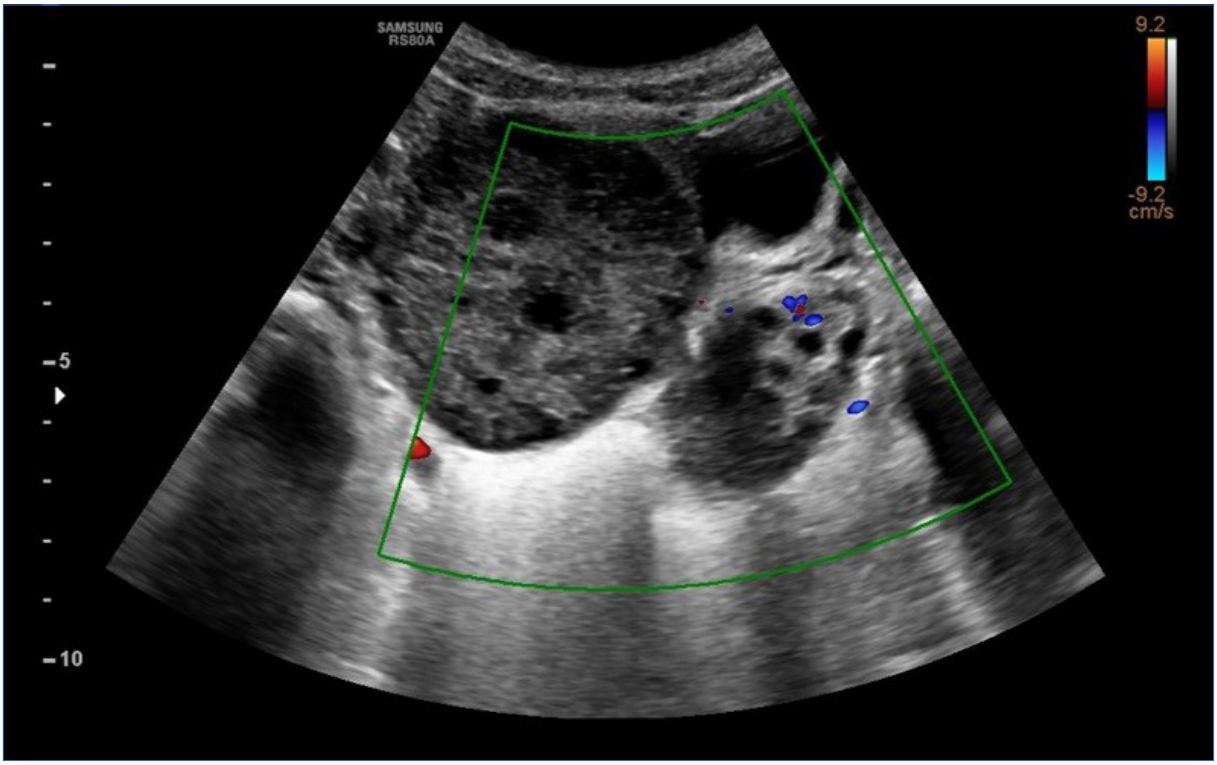

Figure 1. Abdominal ultrasound. Enlarged right ovary with a volume of 185.5 cc (10 x 5 x 6 cm) with a solid, rounded appearance and multiple cysts of up to 15 mm. Left ovary 27.22 cc (2.5 x 4.5 x 3 cm) oval with follicular cysts (<10 mm).

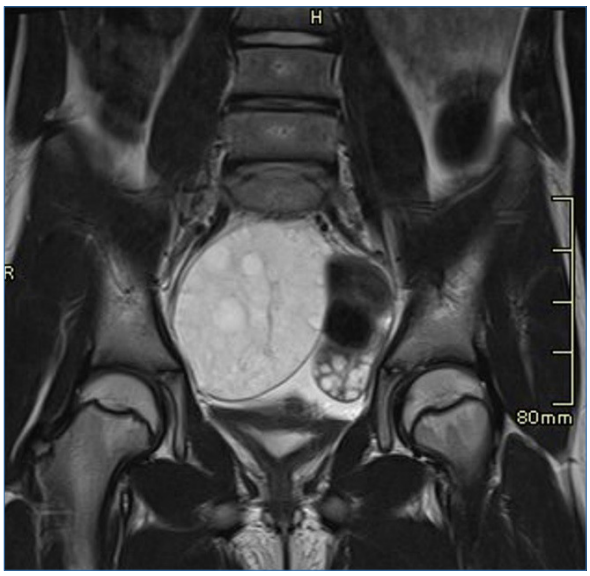

Figure 2. Oedematous right ovary with multiple cysts in its periphery (the largest measures 15 mm) and free fluid in pelvis.

(85 cc) with asymmetric arterial flow compared to the contralateral and ascites. A new intervention was carried out with detorsion (2 turns) and attachment of the adnexa. A control ultrasound was performed at one month without any alterations (right ovary 23.4 cc, left 22.6 cc).

Discussion

Ovarian torsion in girls is a rare pathology that represents a surgical emergency, the most frequent associated symptoms of which are abdominal pain, nausea and vomiting [5,6]. The most frequent radiological findings are asymmetric enlargement of the ovary, peripheral location of the follicles and sometimes coexistence of an ovarian mass inside [6,7]. All these findings were present in our patient.

Most cases of paediatric ovarian torsion occur in ovaries with an adnexal pathology, such as teratomas or cysts. The risk of torsion increases when the mass is benign and large, with a size of 5 centimetres or more [3,8]. Our patient's case is peculiar since she did not present any large mass, only follicular cysts smaller than 1.5 cm.

Surgical treatment options include isolated detorsion, detorsion with oophoropexy, and oophorectomy. In the paediatric population, laparoscopic ovarian detorsion and preservation should be the procedure of choice [3,4,10]. However, abdominal magnetic resonance showed ovarian oedema and free fluid in pelvis. Some authors suggest that oedema can present as a result of partial intermittent torsion of the ovarian pedicle and may present as a solid, adnexal mass [9]. Idiopathic recurrence of ovarian torsion is a phenomenon rarely found in the literature, accounting for around 5-18% of the cases described in different reviews [4]. After idiopathic recurrence, oophoropexy by permanent suture or by plication of the utero-ovarian ligament are the recommended treatments, although this does not completely rule out the possibility of an adnexal torsion in the future [11]. In the case of our patient, her initial oophoropexy did not prevent a subsequent adnexal torsion. Our case also represents a diagnostic challenge regarding the characteristics of precocious puberty present prior to the appearance of the ovarian torsion. In the clinical evaluation, the patient presented symptoms compatible with peripheral precocious puberty with bilateral thelarche and advanced bone age, not mediated by the hypothalamic-pituitary-gonadal axis [1]. The ultrasound scan revealed the presence of enlarged ovaries and multiple cysts, which may lead to the suspected presence of functional cysts, the most frequent cause of peripheral precocious puberty [1,2]. However, hormonal studies are not conclusive of peripheral precocious puberty since we did not find an increase in sex steroids as might be expected [1,2].

Taking into account the presence of intermittent vaginal bleeding and recurrent ovarian torsion, in our patient we can assume the presence of functional ovarian cysts with oestrogen production that may have returned due to associated ischaemic-haemorrhagic phenomena, as has been proposed by other authors [2]. This would also explain why we obtain low estradiol levels when making analytical determinations.

Conclusion

In conclusion, precocious puberty in girls may be due to the presence of follicular cysts with autonomous oestrogen production. On the other hand, the presence of several of these cysts favours the development of ovarian torsion, with some cases of recurrent episodes. These episodes of ovarian torsion can cause ischaemic-haemorrhagic phenomena in the cysts, leading to their autoregression or functional involution. We must be aware of the existence of this phenomenon when studying girls with clear signs of precocious puberty in whom a central origin has been ruled out and who present ovarian cysts and low estradiol levels in blood tests.

Conflicts of interest

The authors declare no conflicts of interest related to this article.

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