CASE REPORT

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Imperforate hymen with hematocolpos. A case report

Himen imperforado con hematocolpos. A propósito de un caso

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ABSTRACT
Imperforate hymen is a rare congenital malformation recognized as the most common cause of hematocolpos. Most patients with imperforate hymen are underdiagnosed until the manifestation of obstructive symptoms in adolescence. We present the case of a 13-year-old female diagnosed with imperforate hymen with hematocolpos.
Key words: Hymen, Hematocolpos, Pelvic pain, Amenorrhea

RESUMEN
El himen imperforado es una malformación congénita poco frecuente reconocida como la causa más común de hematocolpos. La mayoría de las pacientes con himen imperforado son infradiagnosticadas hasta la manifestación de síntomas obstructivos en la adolescencia. Presentamos el caso de una mujer de 13 años a quien se le diagnosticó de himen imperforado con hematocolpos.
Palabras clave. Himen, Hematocolpos, Dolor pélvico, Amenorrea

Introduction

Imperforate hymen is a rare congenital anomaly related to absence of tearing of this membrane during neonatal development[(1)]. Most patients remain asymptomatic until menarche when they begin to present symptoms of obstruction such as cyclic abdominal pain and amenorrhea[(1-3)].

The diagnosis is based on the absence of perforation of the hymenal membrane at the level of the vaginal introitus on pelvic inspection[(1)]. The main clinical manifestation is hematocolpos which consists in the accumulation of blood in the vagina or in the uterine cavity due to the obstruction generated by the absence of perforation of the hymen that prevents the outflow of blood during menstruation. Hematocolpos may manifest with bulging of the hymenal membrane and he presence of a hypogastric mass painful to the touch, which is confirmed by ultrasound[(1-4)].

Treatment is surgical, by opening the hymen and draining the accumulated material. After the intervention, most of the patients remain asymptomatic and start menarche, being reduced the possibility of recurrence[(3)].

Case report

The case of a 13-year-old Arab girl who was referred to the pediatric emergency department for abdominal pain of several weeks' evolution is presented. She was afebrile with no other accompanying symptoms.

The patient had no known drug allergies or other medical history of interest. She referred surgical intervention for strabismus at the age of 8 years and absence of menarche.

On physical examination she showed hypogastric abdominal distension, painful on palpation, with no signs of peritoneal irritation. The ex-

ternal genitalia were normal, but at the level of the vaginal introitus a bulging bluish membrane was visible (Figure 1), with no possibility of vaginal examination by speculoscopy.

Evaluation by abdominal ultrasound showed a regular anteverted uterus with a hysterometry of 6 cm and the endometrium was linear. An echonegative formation measuring 120 x 84 mm was observed at the level of the vaginal cavity (Figure 2). Both adnexae were normal with no evidence of free fluid in the Douglas pouch. With these findings, the diagnosis was imperforate hymen with hematocolpos.

The therapeutic approach consisted of hymenectomy. In the operating room, under general anesthesia a 1 cm longitudinal incision was made at the level of the hymeneal membrane followed by drainage of abundant dark hematic material.

The patient is currently asymptomatic and has regular menstrual cycles, without any adverse event.

Figure 1. External genitalia and vaginal introitus. Bulging and bluish hymenal membrane with absence of perforation is shown.

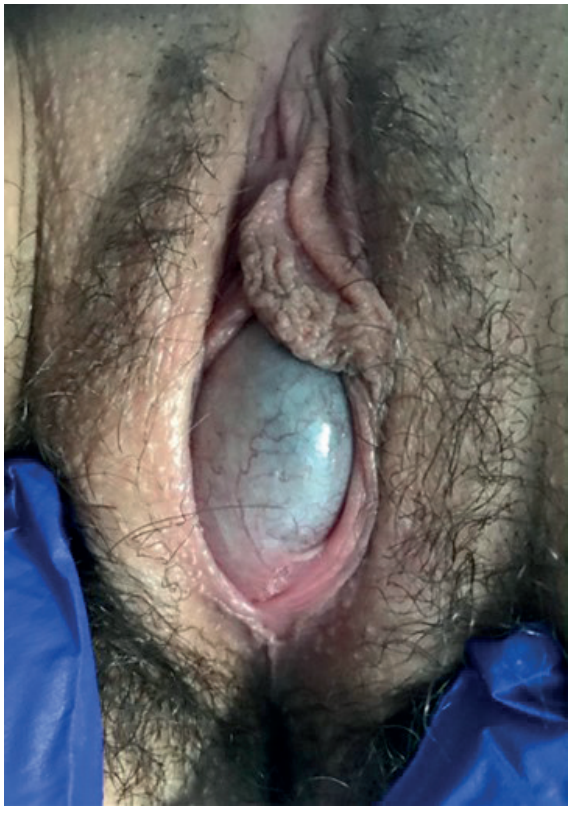
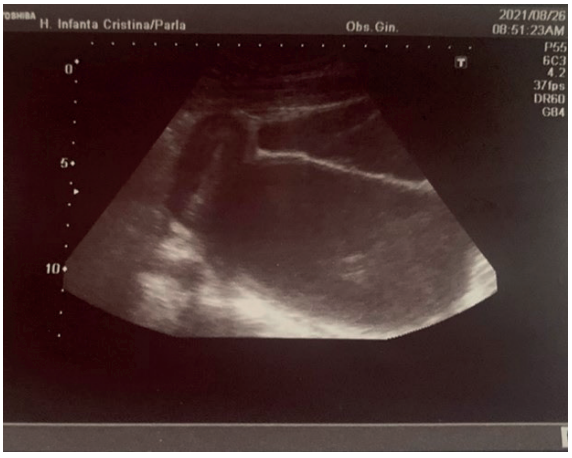


Figure 2. Abdominal ultrasound: regular anteverted uterus with linear endometrium. Anteuterine formation compatible with hematocolpos.



Discussion

Imperforate hymen, despite being a rare disease, is the most frequent congenital anomaly of the female internal genitalia, with an incidence of 1 per 1,000 women, according to the American College of Obstetricians and Gynecologists (ACOG)[5].

It has been associated with amenorrhea and hematocolpos, being the cause of 90% of hematocolpos, but it is the least frequent among the different causes of amenorrhea. Most occur sporadically, although cases associated with autosomal dominant and recessive inheritance have been described[1,2,4].

The absence of hymenal perforation is related to an alteration in the canalization of the vagina and the absence of degeneration of epithelial cells during embryonic development[1,2]. In addition to imperforation, there are other pathological forms of the hymen such as microperforation, cribriform or septate hymen[4,6], being the annular and fimbriated the most common within normal hymens[1].

The form of presentation is variable. Most of the symptoms are derived from the accumulation of blood due to the obstruction generated by the imperforate hymen. In the study by Darwish et al[3], of the 36 patients evaluated with imperforate hymen, 100% presented hematocolpos and 70% had cyclic pelvic pain.

Although rare, associated anomalies at the level of the kidneys, ureters or bladder should be

ruled out, since the development of the genital tract is closely related to the urinary system[6].

Lee K. et al[7] conducted a study in which 236 women with imperforate hymen were included. 27.7% showed urinary retention, while only 5% had renal involvement. Cases of repeated urinary tract infections and urinary incontinence related to accidental penetration of the urethra have been described[3].

In the diagnosis, the importance of a good anamnesis about menarche and coital history is emphasized[1]. Most are diagnosed between 12-18 years of age by examination of the internal genitalia[7], visualizing a bulging bluish membrane covering the entire vaginal introitus[1]. When performing the Credé maneuver, it is possible to observe the increase of the bulge[1].

Although they are usually diagnosed at puberty, symptomatic pediatric cases have been detected and even cases diagnosed by prenatal ultrasound after the finding of hydrocolposis[1,2]. These are generated by accumulation of urogenital secretions under maternal estrogenic influence and most of them end up being reabsorbed[6,8].

The importance of not delaying diagnosis lies in the possibility of developing complications such as hydronephrosis, endometriosis and infertility, in addition to perforation of the hematocolpos[1]. The few published cases of death have been associated with respiratory failure due to abdominal distension[1,2,4].

Abdominal ultrasound is useful in the confirmation of the diagnosis, not requiring any additional test[9]. However, for differential diagnosis, pelvic MRI with anatomical study of the pelvis may be necessary[1].

Vaginal agenesis, partial hymenal obstruction and low transverse vaginal septum are included in the differential diagnosis[5]. The association with McKusick Kaufman syndrome, which also includes polydactyly, congenital heart disease and hydrometrocolpos should be considered[1]. However, most cases occur in isolation, as demonstrated by Lee K. et al[7] who, of the 236 cases studied, 116 had imperforate hymen without other associated pathology.

Partial obstruction of the hymen, which can manifest as septate, cribiform or microperforated hymen, is usually asymptomatic, with consultation due to the impossibility of using a tampon or coital problems. Treatment is surgical, similar to surgery for imperforate hymen, resecting the excess hymenal tissue[6].

Vaginal agenesis consists of the absence of vagina with variable development of the uterus. Although less frequent than imperforate hymen, it is the second most frequent cause of primary amenorrhea after gonadal dysgenesis. On physical examination, the external genitalia are normal, presenting a small concavity at the vaginal level and absence of the hymen. Unlike imperforate hymen, the first-choice treatment recommended by ACOG is the use of vaginal dilators, with excellent results and a high degree of patient satisfaction[6,8].

Transverse vaginal septum consists of the formation of a septum, in 46% of cases at the level of the upper third of the vagina, partially obstructing the vagina and potentially developing hematocolposis. On examination, the septum prevents visualization of the cervix, requiring the use of ultrasound or MRI to distinguish it from agenesis of the cervix. Treatment is surgical, although with worse results than hymenectomy of imperforate hymen, with a higher rate of recurrences and comorbidities such as dyspareunia and dysmenorrhea[6,8].

The treatment of imperforate hymen is surgical. The ACOG recommends cross hymenectomy, which consists of draining the accumulated material through an incision at the level of the hymenal membrane[9].

Several studies have compared different types of surgical approaches, evaluating efficacy and risk of recurrence. They include cross hymenectomy, hymenectomy by electric scalpel, use of the Foley catheter and dilatation by Hegar dilators. There were no statistically significant differences in the number of restenosis depending on the type of technique used. However, the use of topical estrogen creams is discouraged due to a slight increase in recurrences after their use[1].

Some researchers criticize the Foley catheter technique, since maintaining it for at least two weeks increases the risk of local infection[3].

The innovative technique in the study by Darwish et al[3] demonstrated a high satisfaction rate for patients with imperforate hymen. The procedure consisted of performing the hymenal incision with the triangular tip of a 10 mm laparoscopic trocar.

One of the main drawbacks of the treatment is related to the patient's religious customs. However, information should be provided on the possible complications of not performing the operation, and there are serious cases in the literature, such as perforation of the hematocolpos with generalized peritonitis[1,3].

After surgery, the symptoms reported by the patient disappear, with a very low percentage of recurrences. It is estimated that only 6.6% manifest restenosis or vaginal adhesions after surgery[7,8]. Even in cases where infection has been reported after the incision, the evolution is satisfactory. Postoperative comorbidity is low and 86% of patients do not present dyspareunia[1]. It is therefore advisable to carry out a subsequent check-up in order to assess the absence of recurrences and adverse effects.

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