

A didelphic uterus and imperforate hymen presenting as a double hematometrocolpos - a case report

Duplo hematometrocolpos como forma de apresentação de um caso de hímen imperfurado associado a útero didelfos – a propósito de um caso clínico

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Abstract

The rare finding of both a didelphic uterus and imperforate hymen is described in this case. Imperforate hymen is an important cause of abdominal pain in female adolescents, which can usually be diagnosed by thorough clinical history and physical examination. Further investigation may be necessary to exclude other genital tract anomalies.

Keywords: Primary amenorrhea; Abdominal/pelvic pain; Imperforate hymen; Didelphys uterus; Hematometrocolpos.

INTRODUCTION

Müllerian malformations represent a wide spectrum of anomaly of the female reproductive tract, usually detected at birth or at puberty. Didelphys uterus is one of the least common anomalies, representing approximately 5 to 7% of müllerian defects¹. In this condition, there is non obstructed failure of lateral fusion involving both the uterus and vagina.

Imperforate hymen represents the extreme in the spectrum of embryological variations in hymenal configuration, with reported incidence ranging from 0.014% to 0.1%². An imperforate hymen is a congenital resorptive defect which, on the other hand, does not apparently derive from the müllerian ducts. The rare finding of both a didelphys uterus and coexisting imperforate hymen is described in this case of a young patient presenting with pelvic pain and double hematometrocolpos.

CASE REPORT

A premenarchal 13-year-old girl was admitted to the emergency department with severe suprapubic abdominal pain radiating to her lower back of two days' duration. There was no history of nausea, vomiting, altered bowel habits, urinary symptoms or fever. She was apparently asymptomatic until one month before when she was presented to the pediatrics department complaining of lower back pain and was treated as having urinary tract infection. The medical history was unremarkable.

On physical examination we observed normal secondary sexual characteristics. Her abdomen was soft and tender without any palpable mass. Gynecologic examination revealed a bluish bulging hymen. We performed a transabdominal ultrasonography which showed a didelphic uterus (Fig. 1) and a heterogeneous mass in the pelvis measuring about 11 cm. We diagnosed an imperforate hymen with a double hematometrocolpos.

The patient was urgently taken to the operating room and a hymenotomy was performed by a Y-shaped incision. Large quantity of chocolate coloured blood was drained. Intraoperative exam revealed a longitudinal vaginal septum. A few sutures were used to prevent refusion. The postoperative period was uneventful.

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FIGURE 1. Transabdominal ultrasound transverse view showing two hemiuteri

One month after surgery she underwent a magnetic resonance imaging (MRI) which confirmed the existence of a didelphys uterus with two cervix and double vagina (Fig. 2). No urinary tract anomalies were found.

During 3 month follow up, the girl was asymptomatic and had regular normal menstrual cycles. She was sent to a tertiary referral hospital for uterovaginal anomalies management.

DISCUSSION

The presence of a didelphys uterus and concomitant imperforate hymen is likely to be coincidental, since these portions of the female genitals derive from two distinct embryological structures and these defects occur at different stages of embryological development.

Didelphys uterus arises when midline fusion of the müllerian ducts is arrested, either completely or incompletely. The complete form is characterized by 2 hemiuteri, 2 endocervical canals with cervixes fused at the lower uterine segment. Each hemiuteri is associated with one fallopian tube. The vagina may be single or double, with duplication a frequent component. The double vagina manifests as a longitudinal septum that extends either completely (complete septum) or partially (partial septum) from the cervixes to the introitus. A complete longitudinal vaginal septum occurs in 75% of these anomalies, although vaginal septa can

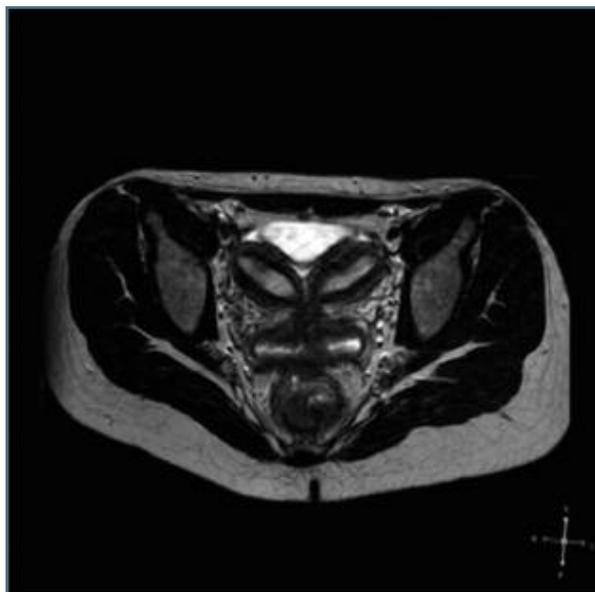


FIGURE 2. Postoperative MRI. Axial T1-weighted image showing didelphys uterus

also coexist with other müllerian duct anomalies^{3,4,5}. Patients with a didelphys uterus are usually asymptomatic, unless an obstruction is present.

The hymen is formed from the endoderm of the urogenital sinus epithelium and represents the junction of the sinovaginal bulbs with the urogenital sinus, becoming perforate before or shortly after birth⁶. An imperforate hymen is the result of failure of canalization of the vaginal plate. The reason for non-perforation of this membrane is unknown.

Imperforate hymen occurs mostly in a sporadic manner, although some familial occurrences have been reported.

It mostly presents during puberty^{7,8} although diagnoses in utero^{9,10,11} and during the newborn period and childhood⁹ are also documented.

Imperforate hymen is a rare though a serious cause of abdominal pain in female adolescents, that may not be detected until the onset of menses, when hematocolpos causes symptoms due to expanding pelvic mass. Hematocolpos can be suspected in adolescent girls in the age group of 13-17 years who present with primary amenorrhea, a cyclic pattern of lower abdominal/pelvic pain, with or without associated symptoms like back pain (38%–40%), urine retention (37%–60%) or constipation (27%)¹¹⁻¹³. The accumulation of menstrual blood in the vagina and uterus may result in a mechanical effect on the urethra, bladder or intestines and

lead to the obstructive urinary or intestinal symptoms¹⁴. Irritation of the sacral plexus or nerve roots is postulated as a mechanism for the referred low back pain¹⁵.

On physical examination, a lower abdominal mass may be palpable, or a pelvic mass may be detected on bimanual rectal examination. Perineal examination reveals a bluish bulging hymen at the introitus. The diagnosis can be established with an abdominal ultrasound showing the pelvic cystic mass.

Other important differential diagnoses of abnormal hymen include vaginal agenesis, transverse vaginal septum and labial adhesion⁷.

Since imperforate hymen is generally considered not to be associated with other Müllerian abnormalities, further investigation of these patients for concomitant urogenital abnormalities has been thought to be unnecessary^{11,16}. However, a few associated anomalies with imperforate hymen have been reported in literature and despite their low prevalence, some authors recommend further investigation to rule them out¹⁷.

Treatment of hematocolpos or hematometra mainly consists of surgical hymenotomy under general anaesthesia which allows the accumulated blood to drain away. Simple vertical, T-shaped, Y-shaped, cruciform and X-shaped may be used. X-shaped and Y-shaped incisions has the advantage of reduced risk of injury to the urethra which should be stented during the procedure.

Early diagnosis of an imperforate hymen is important, since it can lead to serious complications such as infections, hydronephrosis, kidney failure, endometriosis and subfertility¹⁸.

The best approach to management of müllerian ducts anomalies should provide relief of symptoms and preserve reproductive ability. Women with a didelphys uterus and bicollis often have good reproductive outcomes without surgical intervention¹⁹. Resection of the vaginal septum is easy and commonly performed. The decision to perform metroplasty should be individualized, and only selected patients (women with a long history of recurrent spontaneous abortions or pre-term births) may benefit from surgical reconstruction. Referrals to centers with expertise are essential. The surgeon must be experienced with the surgical techniques because the initial procedure is more likely to succeed than follow-up procedures. Reoperation in these cases increases the chance of operative injury and the possibility of a poor functional outcome.

In summary, imperforate hymen is an important cause of abdominal pain in female adolescents, which can usually be diagnosed by thorough history taking and a physical examination. Physicians must be aware of this diagnosis while examining adolescent girls with lower abdominal pain and primary amenorrhea in order avoid delay in the diagnosis and treatment, since late discovery of an imperforate hymen may lead to serious complications. The treatment is surgical and an adequately-performed hymenotomy usually leads to an excellent outcome. As presented in this case further investigation may be necessary to exclude other genital tract anomalies.

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