Abstract

Mullerian anomalies comprise a broad range of anomalies. The clinical syndrome of a double uterus, double cervix with obstruction of the vagina and ipsilateral renal agenesis is rare and a high index of suspicion is necessary to diagnose this disorder. The authors report a case of a 13-year-old girl with progressive abdominal pain during menses seven months after menarche. Magnetic resonance showed a didelphic uterus, right hemihematometrocolpos and suggested the presence of two vaginas. The patient underwent transvaginal excision of the septum with prompt clinical improvement.

Keywords: Mullerian anomalies, Uterus didelphys, Obstructed Hemivagina, Renal agenesis, Herlyn-Werner-Wunderlich syndrome

INTRODUCTION

Uterus didelphys with obstructed hemivagina is an obstructive Mullerian anomaly caused by lateral nonfission of the Mullerian ducts with an asymmetric obstruction. The true incidence of this anomaly is unknown, but has been reported between 0.1% and 3.8%. This condition is almost always associated with renal agenesis ipsilateral to the obstruction, which could be attributable to an embryologic arrest at 8 weeks of gestation that simultaneously affects the Mullerian and the metanephric ducts.

It was first recognized in 1992 and is sometimes referred to as Herly-Werner-Wunderlich (HWW) syndrome. This entity is usually diagnosed at puberty, shortly after menarche. Recurrent and progressive pelvic pain is the main clinical complaint. The clinical presentation may comprise not only a dilated hemivagina, but also a dilated uterine cavity and a fallopian tube, as well as bleeding into the peritoneal space as a consequence of blood stasis and retrograde menstruation in the obstructed system. Occasionally, patients present with fever, peritonitis, purulent vaginal discharge and leukocytosis, leading to a misdiagnosis of Pelvic inflammatory disease (PID). Endometriosis can result from blood reflux into the abdominal cavity and may have dire consequences. A right sided prevalence of the ob-
structured system has been described. An early and accurate diagnosis of this entity is important, so that the resection of the obstructing vaginal septum can provide relief of pain and prevent further complications.

The authors describe a case of a didelphic uterus and obstructed hemivagina with ipsilateral renal agenesis whose initial symptom was vulvar discomfort erroneously diagnosed as vulvovaginitis.

CASE REPORT

A 13-year-old girl presented to our Emergency Department with a 1-month history of perineal pain, dysuria, vulvar discomfort and vaginal discharge. She had been treated with antimycotics, with no improvement, by her General Practitioner. She experienced menarche 7 months before admission, her menstruation cycles were regular but she complained of progressive abdominal pain during menses. A gy-
naecologic examination failed to reveal any anomalies of her external genitalia and a uni-digital vaginal palpation revealed a cystic mass bulging through the anterior vagina wall with 7x4 cm. An abdominopelvic ultrasound examination disclosed the absence of the right kidney and a dilated right hemiuterus, and a magnetic resonance image (MRI) showed a didelphic uterus (Fig.1) and a right hemihematometrocolpos, (Fig.2) suggesting the presence of two vaginas (Fig.3). In the examination carried out under general anaesthesia, the bulging of the vagina was identified as a right transverse vaginal septum (Fig.4) and the left portion of the vagina had its own cervix and was free from obstruction. A transvaginal excision of the septum was performed, about 300 cc of haematic fluid were evacuated (Fig.5) and the right sided cervix was exposed, followed by marsupialization of the vaginal septum (Fig.6). The postoperative course and the long-term follow-up were uneventful.

DISCUSSION

Uterus didelphys with obstructed hemivagina is a rare entity and a high index of suspicion is required to identify this uterine anomaly. In a retrospective case review (n=8) the mean age of diagnosis was 14.5 years with menarche at the mean age of 12.5 years. All the patients had a history of regular menses with cyclic pelvic pain and the mean time to diagnosis was 37.8 weeks\(^2\). In the present case, the correct identification of this situation was one month delayed due to a misdiagnosis of a mycotic vulvovaginitis. Perineal pain or vaginal discharge are not often described as initial symptoms. In this case, digital pelvic examination provided useful information and raised the suspicion of an obstructive mullerian anomaly.

Prior to any surgical intervention, imaging studies should be obtained. The evidence of haematocolpos on an ultrasound, appearing as a fluid collection with low-level echoes, permits an easier detection of the uterine anomaly (didelphic/bicornuate bicollis uterus)\(^3\). It can also confirm the absence of one kidney, which in the presence of an obstructed genital tract should promptly lead to the correct diagnosis\(^2\). MRI is considered to be more sensitive for imaging soft tissue compared with Computerised Tomography (CT), however it is unable to accurately assess the presence of endometriosis, pelvic infection or adhesions that can affect future fertility\(^2\). A few case series confirm that laparoscopy should be considered the gold standard for the complete evaluation of congenital anomalies of the female reproductive...
tract. In our patient, the ultrasound confirmed the presence of two uterine cavities and the absence of the right kidney and the MRI helped characterizing the uterine anomaly.

Currently, the preferred surgical approach for patients with HWW syndrome is the full excision and marsupialization of the vaginal septum. Errors in the surgical management can occur when the diagnosis is not suspected and a laparotomy resection of the intrabdominal mass is attempted. As an alternative to the conventional treatment, a hysteroscopic resection of the vaginal septum under transabdominal ultrasound guidance can be performed. Hysteroscopy considerably enlarges the surgical view which facilitates the preservation of the hymen integrity and the correction of small hematocolpos. In the current case, a classical surgical approach was preferred in order to rapidly relief the perineal pain caused by a tense right hematocolpos.

The present case demonstrates that despite the rarity of this entity it was possible to ascertain the correct diagnosis and, therefore, prevent complications and consequently preserve fertility.

REFERENCES