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OHVIRA-syndrome (Obstructed hemivagina with ipsilateral renal anomaly) as differential diagnosis of acute lower abdominal pain

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OHVIRA-syndrome (Obstructed hemivagina with ipsilateral renal anomaly) as differential diagnosis of acute lower abdominal pain

INTRODUCTION: *OHVIRA-syndrome (obstructed hemivagina, ipsilateral renal agenesis/anomaly) is a rare Mullerian duct anomaly that can lead to complications in pubescent children.*
CASE REPORT: *We report a case of a 13-year-old patient with acute right-sided lower quadrant abdominal pain who was referred for exclusion of appendicitis. As a result of the examination (transvaginal ultrasound scan and gynecological examination), a female genital tract anomaly was suspected in the form of obstructed hemivagina with hematocolpos and hematometra. The MRI scan showed hematocolpos and hematometra on the right side, uterus didelphys accompanied by right-sided renal agenesis, consistent with OHVIRA-syndrome. Excision of the vaginal septum was performed and the accumulated old menstrual blood, as represented by hematocolpos and hematometra, was evacuated. Postoperative recovery was uneventful.*
CONCLUSION: *The early surgical management of this rare Mullerian duct anomaly is important in order to prevent long-term complications. This malformation should be considered in the differential diagnosis of acute lower abdominal pain in pubescent girls.*

KEY WORDS: Abdominal Pain, Genital Anomaly, Obstructed Hemivagina, Renal Anomaly

Introduction

Mullerian malformations have an incidence of 5,5%, of which 36% occur with other structural abnormalities and most of these are renal anomalies [1-5].
The OHVIRA-syndrome (obstructed hemivagina, ipsilateral renal agenesis/anomaly) has an unknown incidence and is a rare form of defective fusion of the Mullerian ducts during the development of the female reproductive system. This Mullerian anomaly is characterized by obstructed hemivagina, uterine anomaly and ipsilateral renal anomaly. It can be associated with other anomalies such as intestinal malrotation [6].
The uterine malformations have an estimated incidence of 7% to 10% in the general female population [7]. This syndrome remains asymptomatic in prepubescent children, the first symptoms begin after the menarche because of the disturbance of the normal physiological process [2,4,8]. Clinical symptoms vary widely, from cyclic, lower abdominal pain resulting from retained menstrual blood in the obstructed hemivagina, to urinary symptoms and infertility [8-10]. The acute or chronic lower pelvic pain occurred shortly after menarche, secondary to hematocolpos [7].
An accurate diagnosis is important and can be done using ultrasound, computed tomography, magnetic resonance imaging, and direct visualization via hysteroscopy or laparoscopy. The delay or lack of therapy may lead to an increased risk of complications like endometriosis, pelvic adhesions, pyometra and infertility [8-11].
The surgical management of the OHVIRA-syndrome is recommended as soon as possible in order to reduce the

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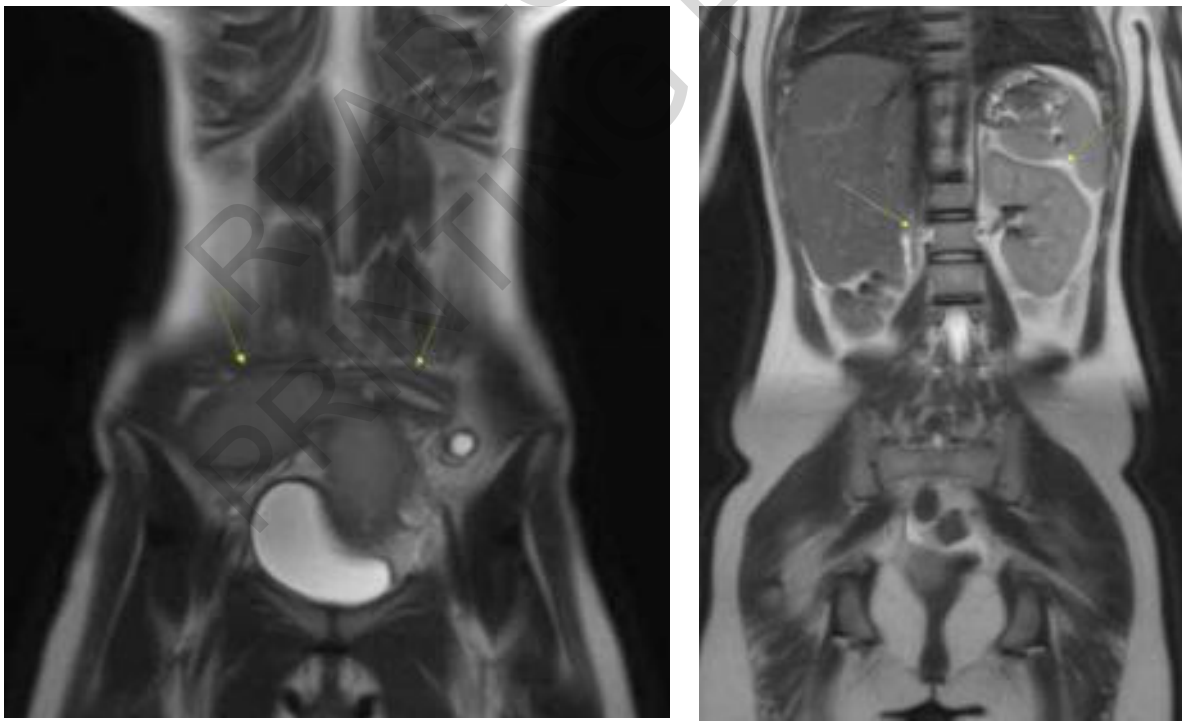
long-term complications resulting from the retrograde flow of menstrual blood [9]. The surgical management of the OHVIRA-syndrome and the ability to maintain hymenal integrity in pubescent patients play a very important role, considering the varying cultural norms surrounding hymenal integrity [9].
A good prognosis is achieved through an early diagnosis and adequate therapy [11]. Long-term follow-up of the patients after the therapy of OHVIRA-syndrome reduces complications like dyspareunia, vaginal stenosis and recurrence of the obstruction [11].

Case Report

A 13-year-old patient with acute right-sided lower quadrant abdominal pain, severe tenderness over McBurney's point and palpable pelvic mass was referred for exclusion of appendicitis.
Case history: menarche at the age of 11, regular menstrual cycles, dysmenorrhoea in the last three months, sexually active, attested mental retardation. No previous history of operations. The family history was unremarkable. There were no reported problems in micturition and defecation, as well as no nicotine or alcohol abuse. The transvaginal ultrasound scan showed an elongated, hypoechoic mass in the right iliac fossa, while the uterus and adnexa were deviated to the left.
The gynecological examination revealed normal external genitalia, open vaginal introitus, no hymenal atresia, protrusion of the right livid vaginal wall, and the ectocervix was deviated to the left. As a result of the examination, a female genital tract anomaly was suspected in the form of obstructed hemivagina with hematocolpos and hematometra.
For further accurate and comprehensive diagnosis, a magnetic resonance imaging scan (MRI) was performed. The MRI scan showed hematocolpos and hematometra on the right side and uterus didelphys accompanied by right-sided renal agenesis, consistent with OHVIRA-syndrome (Fig. 1).
After the preoperative preparation, a scalpel excision of the thin vaginal septum was performed and the accumulated old menstrual blood, in the form of hematocolpos and hematometra on the right side, was evacuated. The right side of the cervix was not palpable, it was probably dilated due to the accumulated blood. Postoperative recovery was uneventful.

Discussion and Conclusion

The early diagnosis and proper therapy of this malformation, based on invasive methods (laparotomy, vaginal surgery, laparoscopy, hysteroscopy) and non-invasive methods (two- or three-dimensional transvaginal ultra-



A) B)

Fig. 1: A) Preoperative image of a right-sided hematocolpos and hematometra, uterus didelphys. B) Uterus didelphys accompanied by right-sided renal agenesis.

sound, MRI, three-dimensional saline infusion sonovaginocervicography) play a key role [3,12].

Based on the MRI's high accuracy rate in diagnosing congenital uterine anomalies and the simultaneous assessment of the cavity and fundus of the uterus, we used the classic methods of diagnosis and therapy, however, three-dimensional ultrasound is preferred by some clinicians in the diagnosis of the congenital uterine anomalies [3,12]. In our case, virginity-sparing management was not an issue, but some authors reported the good results of septoplasty supported by the use of three-dimensional saline infusion sonovaginocervicography with virtual speculoscopy and minimally invasive surgery (for example hysteroscopic resection) as a virginity-sparing management option [1,2,4,12].

The early surgical management of this rare Mullerian duct anomaly is important in order to prevent long-term complications.

This malformation should be considered in the differential diagnosis of acute lower abdominal pain in pubescent girls.

Riassunto

Introduzione. La sindrome OHVIRA (emivagina ostruita, agenesia/anomalia renale omolaterale) è una rara anomalia del dotto mulleriano che può portare a complicazioni nelle bambine all'epoca della pubertà.

Caso clinico. Riportiamo il caso di una ragazza di 13 anni con dolore addominale acuto al quadrante inferiore destro che è stato inviato con diagnosi di probabilità di appendicite. A seguito dell'esame (ecografia transvaginale ed esame ginecologico) è stata sospettata un'anomalia del tratto genitale femminile sotto forma di emivagina ostruita con ematocolpo ed ematometra. La risonanza magnetica ha mostrato ematocolpo ed ematometra sul lato destro, utero didelphys accompagnato da agenesia renale sul lato destro, compatibile con la sindrome OHVIRA. È stata eseguita l'asportazione del setto vaginale ed è stato evacuato il vecchio sangue mestruale accumulato, rappresentato dall'ematocolpo e dall'ematometra. Il recupero postoperatorio è stato regolare.

Conclusione. La gestione chirurgica precoce di questa rara anomalia del dotto mulleriano è importante per prevenire complicazioni a lungo termine. Questa malformazione dovrebbe essere considerata nella diagnosi differenziale del dolore addominale inferiore acuto nelle ragazze pubescenti.

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