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Herlyn Werner Wunderlich Syndrome with Hematocolpos Symptom

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Abstract

BACKGROUND: Uterodidelphys with obstructed hemivagina and ipsilateral renal agenesis is referred to as the Herlyn Werner Wunderlich (HWW) syndrome. Herlyn-Werner-Wunderlich (HWW) syndrome is a very rare congenital anomaly of the urogenital tract involving Müllerian ducts and Wolffian structures, and it is Werner by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis.

CASE REPORT: Here, we describe a 17-year-old female patient presented with severe and increasing cyclical abdominal pain. She attained menarche 3 years back. The patient had irregular and scanty menstruation associated with dysmenorrhoea. On physical examination, secondary sexual characters were well developed, and on palpation, lower abdomen tenderness was not present. Diagnosed with HWW syndrome, who was taken up for diagnostic sonography and MRI, followed by vaginal septal resection,

CONCLUSION: In addition to a definitive diagnosis, this approach helped in symptomatic relief to the patient.

Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome is a very rare congenital anomaly of the urogenital tract involving Müllerian ducts and Wolffian structures, and it is Wunderlich by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis [1], [2].

Combination of obstructed hemivagina and uterus didelphys was first reported in 1922 [3]. The triad was reported in 1971 by Herlyn and Werner and again in 1976 by Wunderlich. The incidence of didelphys uterus, related to HWW, is approximately 1 / 2,000 to 1 / 28,000, and it is accompanied by unilateral renal agenesis in 43% of cases. The incidence of unilateral renal agenesis is 1 / 1,100, and 25-50% of affected women exhibits associated genital abnormalities [2]. A complete or partial vaginal septum is present in 75% of women with didelphys uterus [1]. The exact cause, pathogenesis and embryologic origin of HWW syndrome are unclear and remain a subject of discussion [3].

Case Report

Seventeen years old female patient presented with severe and increasing cyclical abdominal pain. She attained menarche 3 years back. The patient had irregular and scanty menstruation associated with dysmenorrhoea. On physical examination, secondary sexual characters were well developed, and on palpation, lower abdomen tenderness was not

present.

Routine investigations were normal. From vaginal inspection, it was found that vaginal introitus was seen and with an intact hymen. Rectal examination shows that USG revealed the absence of the right kidney and a uterine didelphys with hematocolpos, and both ovaries were normal. MRI indicated a uterus didelphys with imperforate hymen and hematocolpos on the right vagina (Figure 1).

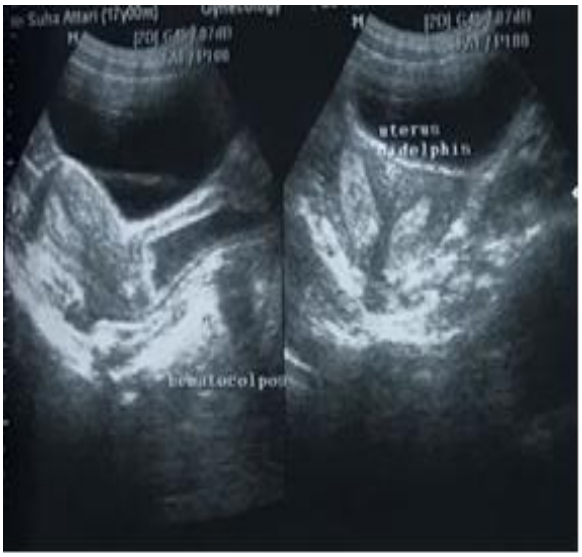

Figure 1: USG showing uterus didelphys + hematocolpos

There was a collection of fluid both in the right uterus and right obstructed vagina suggestive of hematocolpos. With the conclusion of MRI is, uterus didelphys with hematocolpos on the right ovary. BNO-IVP revealed that the right kidney was atresia, but left kidney appeared normal and no sign of obstruction on the left kidney and ureter (Figure 2).

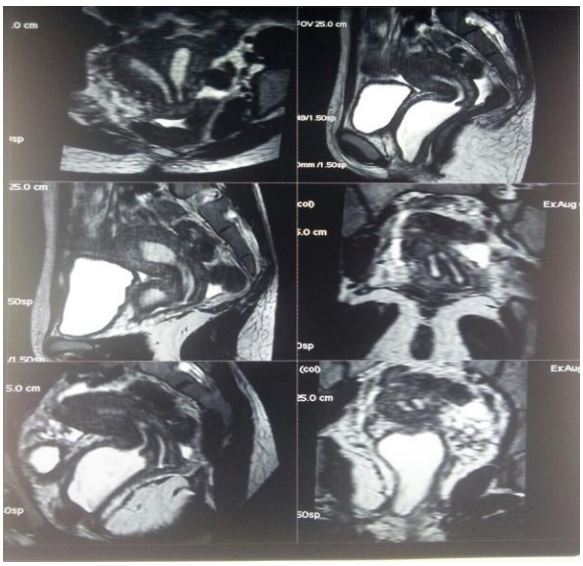

Figure 2: MRI showing the absence of uterus didelphys + hematocolpos

Discussion

Mullerian (paramesonephric) duct anomalies are congenital anomalies of the female genital tract which result from non-development or non-fusion of the Mullerian ducts or failed resorption of the uterine septum during the sixth to ninth weeks of fetal life causing a wide-ranging series of reproductive duct malformations [1], [2].

The paramesonephric ducts of the genetically female embryo fuse in the midline and from the uterus, cervix and the upper four-fifths of the vagina. The lower 20% of the vagina is formed from sinovaginal bulbs which are protrusions of the urogenital sinus [3]. The urinary and genital systems arise from a common ridge of mesoderm arising along the dorsal body wall and rely on the normal development of the mesonephric system. Hence, abnormal differentiation of the mesonephric and paramesonephric ducts may also be associated with anomalies of the kidneys [4]. Renal agenesis is the most common anomaly although horseshoe or pelvic kidney, cystic renal dysplasia, duplication of the collecting system and ectopic ureters have all been described [4], [5].

Renal agenesis is predictive of an ipsilateral obstructive Mullerian anomaly greater than 50% of the time. These anomalies have a right-sided dominance, twice as often as on the left side. Such a relationship between female genital and urogenital anomalies should lead us to examine the urogenital system when a genital anomaly is identified and vice-versa [6], [7], [8].

HWW syndrome is usually discovered at puberty with non-specific symptoms, like increasing pelvic pain, dysmenorrhea and palpable mass due to the associated haematocolpos or hematometra, which result from retained, longstanding menstrual flow in the obstructed vagina. A right-sided prevalence has been described. It is postulated that the right side is more susceptible to hypoxic damage than the left side due to a precocious mitochondrial maturity on the left side, resulting in less tissue damage following hypoxia [6], [7].

If treatment is delayed, complications may develop, such as endometriosis caused by retrograde menstruation, infections and pelvic adhesions, which in turn might obstruct the genital organs. Clinical suspicion and awareness of the syndrome are therefore imperative to making a timely diagnosis and preventing these complications [4], [8].

CT and ultrasound are the most widely used diagnostic tools [9], [10]. However, MRI is considered to be more sensitive for imaging soft-tissue anatomy and delineating subtle findings seen in congenital anomalies. Hence, it should be obtained before any surgical intervention [7]. Laparoscopy is not mandatory but could help confirm the diagnosis when radiologic imaging is inconclusive, especially in those

cases with endometriomas warranting resection [11].

As obstructive genital lesions may be associated with other anomalies such as coarctation of the aorta, atrial septal defects and abnormalities of the lumbar spine, a complete physical examination and abdominal tests may be indicated [8]. Resection of the vaginal septum is the treatment of choice of obstructed hemivagina [4].

Women with uterus didelphys have a reasonable chance of getting pregnant, but the abortion rate is high (74%), and premature delivery is common (22%) [1], [6]. A caesarean section is required in 82% [11]. Evaluation of the genital tract using MRI scanning is recommended in all girls with known renal abnormalities detected antenatally or after that, before the onset of menstruation. This enables us to diagnose some patients before menarche and carry out a surgical correction of the obstruction before any damage has occurred because of haematocolpos, haematometra and retrograde menstruation [4].

In conclusion, the prompt and accurate diagnosis of female reproductive tract disorders, including HWW syndrome is necessary to prevent complications and preserve future fertility. Early recognition of this relatively rare syndrome would lead to the immediate, proper surgical intervention.

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