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Case Report

Herlyn-Werner-Wunderlich Syndrome: A Rare Congenital Uterine Anomaly

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ARTICLE INFO

Article history:

Received: 29 September, 2021

Accepted: 16 October, 2021

Published: 19 November, 2021

Keywords:

Herlyn-Werner-Wunderlich
syndrome

rare

congenital

uterine

anomaly

treatment

ABSTRACT

Introduction: Herlyn-Werner-Wunderlich Syndrome is a rare anomaly and exact incidence is not known. It is usually diagnosed in young girls who present with cyclical abdominal pain along with cyclical menstruation.

Case Report: We present case of two girls who had this syndrome. An ultrasound was performed in both patients. In first patient, it revealed uterus didelphys with hematocolpos on right side. In the second patient, a large hematometra of size 9.8×12.2 cm, along with hematocolpos was present on the left side. Both the patients underwent examination under anaesthesia and excision of septum. They remained asymptomatic during one year follow-up.

Conclusion: Treatment of such cases is excision of septum and to maintain the patency of the outflow tract and prevent stricture formation later. A good follow-up in immediate post-operative period is necessary.

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Introduction

Herlyn-Werner-Wunderlich Syndrome is a rare uterine anomaly usually diagnosed in adolescent girls. It is characterized by uterus didelphys with blind hemivagina and ipsilateral renal agenesis. This case study reports two young girls who were diagnosed with this syndrome and managed subsequently.

Case Report

A 13-year-old girl presented to the OPD with complaints of severe dysmenorrhea for three months. The pain used to occur in the lower abdomen, and she had to take analgesics for pain relief. She was treated by private practitioners on lines of primary dysmenorrhea. Another young girl, 16-year-old, came to OPD with complaints of constant, dull aching pain in abdomen and a small lump in lower abdomen. Initially the pain was associated with menstruation but later on, there was a constant pain that aggravated during her menses.

The first patient attained her menarche five months prior to presenting to the hospital. The first two cycles were relatively painless but after this she started having severe dysmenorrhea. There was no delay in development of secondary sexual characters. The second girl had attained her menarche three years back. Her symptoms also commenced after few cycles and aggravated with each cycle. She was advised an ultrasound by a practitioner which reported a large cystic collection adjacent to the uterus. The ultrasonologist could not make out the origin of the cystic structure.

The secondary sexual characters were normal for age in both the patients. Examination findings revealed nothing significant in first patient but there was a lump arising from pelvis, corresponding to sixteen weeks sized uterus, in second patient. It was a firm, mobile and tender mass. On rectal examination, a bulge was felt on the anterior rectal wall. Apart from this, no other significant findings could be appreciated on clinical examination. An ultrasound was performed in both patients. In first patient, it revealed uterus didelphys with hematocolpos on right side. In the second patient, a large hematometra of size 9.8×12.2 cm, along with hematocolpos was present on the left side. Hence, both the patients were

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diagnosed with mullerian anomaly. MRI was done in both cases which confirmed the findings of ultrasound and, in addition, showed a longitudinal vaginal septum with blind right hemivagina. In first patient right kidney was absent and left kidney was normal whereas in the second one left kidney was absent. Hence, the renal agenesis was on the side of obstruction in the outflow tract in both the patients (Figures 1 & 2).

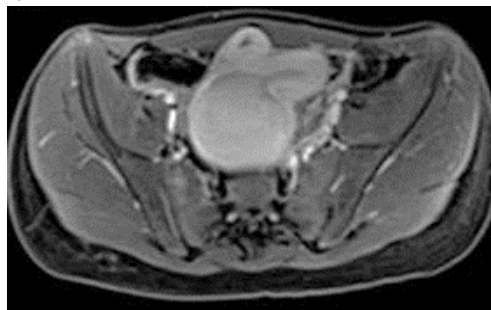


Figure 1: MRI showing uterus didelphys with right sided obstruction leading to formation of right hematocolpos (first case).



Figure 2: MRI showing left renal agenesis in a patient with left outflow tract obstruction (second case).

Both the patients were planned for examination under anaesthesia and excision of septum. In first patient only a cruciate incision could be given over the blind pouch due to lack of space for proper excision to be done. Hematocolpos was drained. Both the cervixes were visualized by a hysteroscope, and they were normal. In post operative period the dilatation of the opening was done by one of the authors by gloved little finger for few days. Later on, patient's mother was taught to perform this maneuver and patient was discharged. Mother was asked to continue this for two weeks to prevent stenosis of the neostoma. Patient was asymptomatic during her one-year follow-up. In the second patient, the septum could be excised and hematometra drained. Two weeks later a one finger vaginal examination was done to evaluate the patency of outflow tract and it was found to be patent. She was also asymptomatic during her eight-month follow-up.

Discussion

Herlyn-Werner-Wunderlich Syndrome is a rare anomaly and exact incidence is not known [1, 2]. The incidence of uterus didelphys, which

is a component of this syndrome, is about 1/2,000 to 1/28,000 which is accompanied by unilateral renal agenesis in 43% of cases [3]. The exact cause, pathogenesis, and embryologic origin of HWW syndrome are not known yet [4]. Patients usually present with dysmenorrhea soon after menarche with normal menstrual flow. They may also present with abdominal or vaginal lump, infertility, and sometimes as acute abdomen [1, 5]. 20% of these patients are diagnosed in their 20s and 10% are diagnosed beyond age 30 years, but most often the diagnosis is made in adolescence [2]. Therefore, there should be suspicion of this syndrome if an adolescent girl presents with these symptoms. Diagnosis can be made by ultrasonography [6]. But along with a pelvic ultrasonographic evaluation, whole abdomen needs to be evaluated for the kidneys otherwise the complete diagnosis could not be made as it happened in this case. MRI is considered to be the modality of choice [7]. MRI is more sensitive in detecting the uterine contour, the shape of the intrauterine cavity and the character of the septum compared to the other modalities. Treatment in these cases is excision of septum and to maintain the patency of the outflow tract and prevent stricture formation. Marsupialization can also be done to prevent stricture of the outflow tract [4]. Early diagnosis of such cases is important to prevent further complications and preservation of fertility. Renal agenesis can be associated with this anomaly. Hence prepubertal girls with renal agenesis should be screened for Herlyn-Werner-Wunderlich Syndrome.

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