Case Report

Herlyn-Werner-Wunderlich syndrome

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ABSTRACT

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), also known as Herlyn–Werner–Wunderlich syndrome, is a rare syndrome constituting 0.16–3% of all the mullerian duct anomalies. Patients are symptomatic shortly after menarche when hematocolpos develop during menstruation, resulting in dysmenorrhea and a pelvic mass. Ultrasonography and magnetic resonance imaging (MRI) are the usual investigations for the diagnosis. Vaginoplasty with septum resection is the treatment of choice to prevent complications such as endometriosis or adhesions from chronic infections with subsequent infertility. Here, we present a case of 13-year-old girl with similar presentation during her second menses and who was symptomatically relieved by vaginal septostomy with marsupialisation.

Key words: Didelphic uterus; Herlyn–Werner–Wunderlich syndrome; mullerian duct anomalies; Obstructed hemivagina and ipsilateral renal anomaly.

Introduction

Herlyn–Werner–Wunderlich (HWW) syndrome is a rare entity, requiring a high index of suspicion for early diagnosis and management. Usually presenting at puberty with dysmenorrhoea during normal menstruation, diagnosis is often delayed due to lack of understanding of this condition by radiologists, gynecologists, and pediatricians.[1] Since this condition can be managed by a simple vaginal septostomy, early diagnosis is mandatory to prevent long-term complications like endometriosis.[2]

Case History

A 13-year-old girl had attended gynecology outpatient with pain abdomen for 25 days along with nausea and intermittent vomiting, which was first such episode. She had attained menarche the previous month with scanty menstrual flow. There was neither history of any medical or surgical disorders nor any significant family history of tuberculosis, or any gynecological disorder including mullerian anomaly. She had been treated with norethisterone for 20 days from outside without any apreciable benefit. There was mild pallor, the uterus was of 14–16 weeks size, and in addition a firm oblong nontender mass was felt on the left illiac fossa arising from the pelvis that was attached to uterus with restricted mobility. Vaginal examination revelaed a nontender cystic bulge in the left lateral vaginal wall. All other systemic examinations were normal.

Ultrasonography (USG) revealed absent left kidney at usual site, and uterus was didelphic type with hematocolpos, hematometra, and hematosalpinx on left uteri and tube with a probable longitudinal septum [Figure 1]. MRI abdomen pelvis was done to confirm USG findings which showed similar findings with absent left kidney [Figure 2].

She was planned for a vaginal septostomy. Cardiology evaluation was normal. Computed tomography urogram was

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done showing absent left kidney and ureter with normal right counterpart. Right cervix was visualized. During septostomy, around 300 ml altered blood was drained [Figure 3]. Intravenous antibiotics was given postoperatively, and the patient did well. After discharge she had normal menses without pain and is still on follow-up without any complaints. Follow-up scans showed normal uteri with resolving hematosalpinx.

Discussion

Mullerian duct anomalies with an incidence of 2–3% constitute an enormous group of complex uterine malformations with or without renal anomalies. HWW syndrome constitutes 0.16-10% of all mullerian duct abnormalities.[2] There are only few few case reports in literature since 1922 and true incidence is still unknown. This triad of didelphic uterus, obstructed hemivagina, and ipsilateral hemivagina was initially disclosed in an English report published in 2006.[3]

Uterus didelphys with obstructed hemivagina is a class III mullerian anomaly having two uteri, two endonetrial cavities, and two cervices. It occurs due to lateral nonfusion of the mullerian ducts with asymmetric obstruction, and it is almost always associated with renal agenesis ipsilateral to the side of obstruction.[4] This anomaly is rarely seen with septate uterus also.[5] A septate vagina occurs in 75% cases. When renal agenesis is encountered first, an ipsilateral obstructive mullerian anomaly is associated about 50% of the time. Apart from ipsilateral renal agenesis, reports of duplicated kidneys, dysplastic kidneys, rectovesical bands, or crossed fused ectopia have also been described.[6] An ectopic ureter may also be encountered which inserts into the obstructed hemivagina and malignant transformation occurring in the ectopic ureter has also been reported.[7]

These patients present with pelvic pain, progressive dysmenorrhea, and an associated pelvic mass, as in our case. But occasionally urgency, frequency, or vaginal discharge may be the presenting symptom.[7] It usually presents at puberty but rarely can present in neonates or in adulthood.[8,9] Physical examination typically reveals a unilateral pelvic mass, more commonly on the right than the left, with a ratio of 2:1.[8] Our patient had left pelvic mass. These obstructive lesions may also be associated with other defects like coarctation of the aorta, atrial septal defects, Inferior vena cava (IVC) duplication, intestinal malrotation, ovarian malposition, and abnormalities of the lumbar spine, thus mandating a complete physical examination with abdominal tests.[10]

USG and MRI are the modalities of choice for the diagnosis and surgical planning of HWW syndrome. Although USG can diagnose this condition, MRI plays an important role in further characterizing the didelphic uterus, obstructed hemivagina, and ipsilateral renal agenesis.[1] Laparoscopy remains the gold standard diagnostic modality for uterine anomalies with the additional therapeutic advantages.

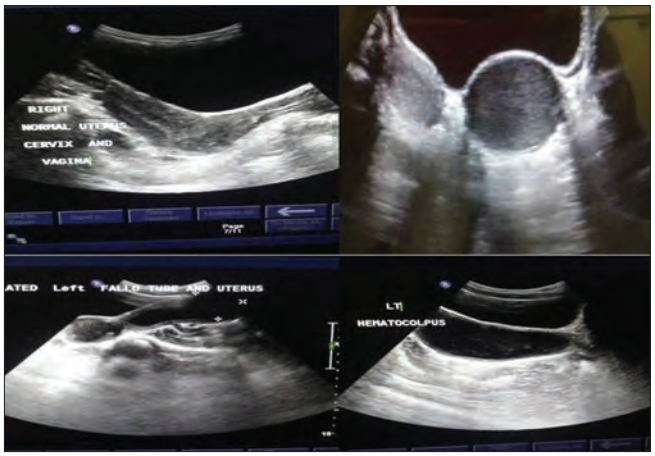

Figure 1: USG findings

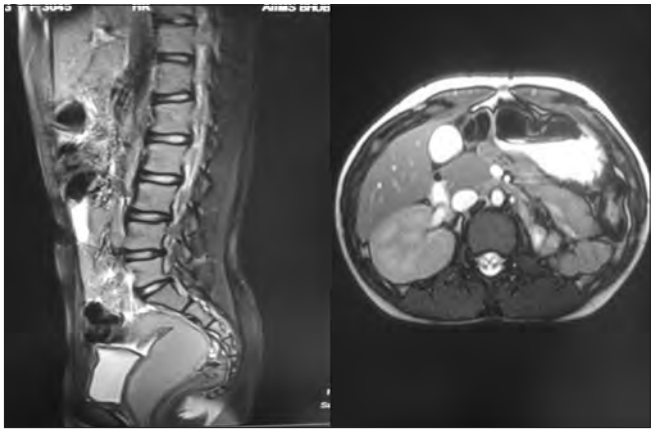

Figure 2: MRI findings

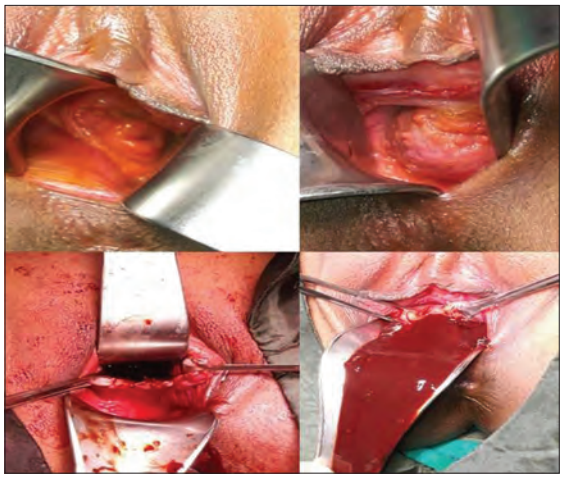

Figure 3: Intraoperative findings

If not treated early then this syndrome can result in serious acute complications like pyohematocolpos, pyosalpinx, or pelviperitonitis, and long-term complications, such as endometriosis, pelvic adhesions, and increased risk of abortion or infertility.[8] Vaginal septum excision is the treatment of choice for HWW syndrome,[10] as done in our case with successful relief of symptoms and restoration of anatomy. With proper management long-term sequelae can be prevented and also successful pregnancy can be achieved in around 87% patients, though 23% have the risk of abortions.[9]

To conclude, HWW syndrome though being a rare entity can be managed well with high index of suspicion and by increasing awareness of this anomaly among health care providers. In prenatal period or during puberty, renal agenesis should raise an alarm of coexistent uterine anomaly and management should be directed toward same to help reducing the morbidities associated with this syndrome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Cox D, Ching BH. Herlyn-Werner-Wunderlich syndrome: A rare presentation with pyocolpos. J Radiol Case Rep 2012;6:9-15.
2. Adair L II, Georgiades M, Osborne R, Ng T. Uterus didelphys with unilateral distal vaginal agenesis and ipsilateral renal agenesis: Common presentation of an unusual variation. J Radiol Case Rep 2011;5:1-8.
3. Gholoum S, Puligandla PS, Hui T, Su W, Quiros E, Laberge JM. Management and outcome of patients with combined vaginal septum, bifid uterus and ipsilateral renal agenesis (Herlin-Werner-Wunderlich syndrome). J Pediatr Surg 2006;41:987-92.
4. Prada Arias M, Muguerza Vellibre R, Montero Sánchez M, Vázquez Castelo JL, Arias González M, Rodríguez Costa A. Uterus didelphys with obstructed hemivagina and multicystic dysplastic kidney. Eur J Pediatr Surg 2005;15:441-5.
5. Haddad B, Barranger E, Paniel BJ. Blind hemivagina: Longterm followup and reproductive performance in 42 cases. Hum Reprod 1999;14:1962-4.
6. Shavell VI, Montgomery SE, Johnson SC, Diamond MP, Berman JM. Complete septate uterus, obstructed hemivagina, and ipsilateral renal anomaly: Pregnancy course complicated by a rare urogenital anomaly. Arch Gynecol Obstet 2009;280:449-52.
7. Boram H, Herndon C, Rosen M, Wang ZJ, Daldrup-Link H. Uterine didelphys associated with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome. Radiol Case Rep 2015;5:327.
8. Wu TH, Wu TT, Ng YY, Ng SC, Su PH, Chen JY, *et al*. Herlyn-Werner-Wunderlich Syndrome consisting ofuterine didelphys, obstructed hemivagina and ipsilateral renal agenesis in a newborn. Pediatr Neonatol 2012;53:68-71.
9. Ahmad Z, Goyal A, Das CJ, Deka D, Sharma R. Herlyn-Werner-Wunderlich syndrome presenting with infertility: Role of MRI in diagnosis. Indian J Radiol Imaging 2013;23:243-6.
10. Aveiro AC, Miranda V, Cabral AJ, Nunes S, Paulo F, Freitas C. Herlyn-Werner-Wunderlich syndrome: A rare cause of pelvic pain in adolescent girls. BMJ Case Rep 2011;2011:bcr0420114147.