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Incidental diagnosis of Herlyn-Werner-Wunderlich Syndrome in a nulliparous woman: a case report in Bhutan

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ABSTRACT

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital syndrome with features of uterus didelphys, ipsilateral absent kidney and obstructed hemivagina. Ultrasound findings of absent kidney or abnormalities in the kidney should alert the clinicians about the syndrome. Complications like endometriosis, infertility and pelvic inflammation occurs with late diagnosis.A 42-year-old nulliparious married woman who presented with right sided abdominal pain to the National Referral Hospital was diagnosed to have HWWS. The diagnosis was supported by ultrasound, CT and MRI findings. Patient was treated for endometriosis and had symptomatic improvement. The diagnosis of the syndrome is challenging as it is rare but clinicians should suspect the syndrome in women who present with infertility. Ultrasound scan is the basic investigation to screen the syndrome which is available in most hospitals.

Keywords: Case report; Herlyn-Werner-Wunderlich syndrome.

INTRODUCTION

Herlyn-Werner-Wunderlich syndrome (HWWS), a Mullerian anomaly has an incidence of 0.1-3.8%[1] The diagnosis is aided by ultrasound, CT or MRI scans. The treatment depends on the variants of the syndrome whether it is class 1 or class 2[2,3].

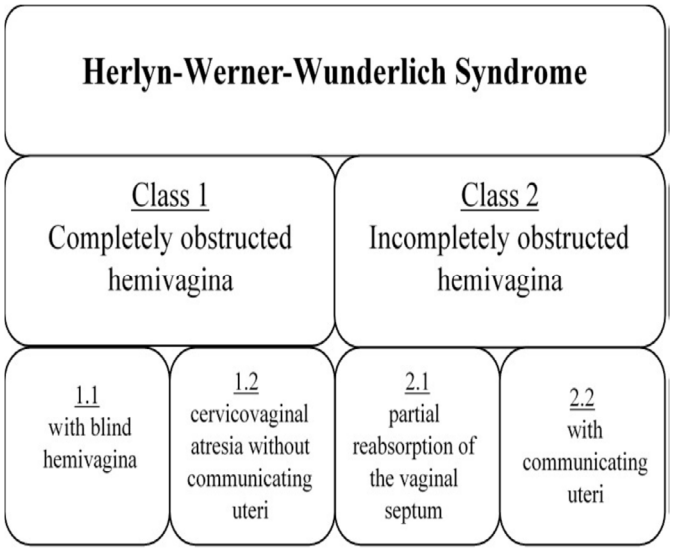

Figure 1. Classification of HWWS based on vaginal morphology

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Herein, we report a case of HWWS incidentally detected in a 42-year-old nulliparous woman with the aim of creating awareness among radiologists, gynaecologists, and general practitioners about the embryology, symptoms, signs and radiological features of HWWS.

CASE REPORT

A 42-year-old married nulliparous woman presented with pain on the right side of the lower abdomen to the National Referral

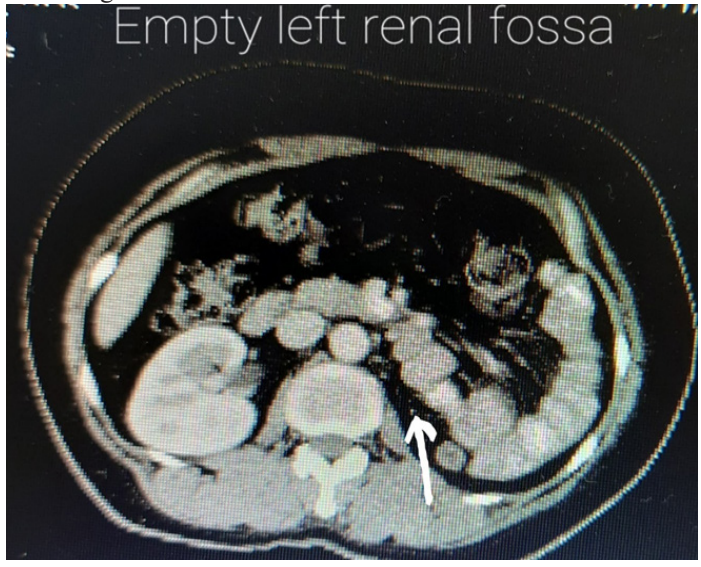

Figure 2. Axial contrast enhanced CT image of the abdomen showing absence of left kidney and renal fossa is occupied by the bowel loops

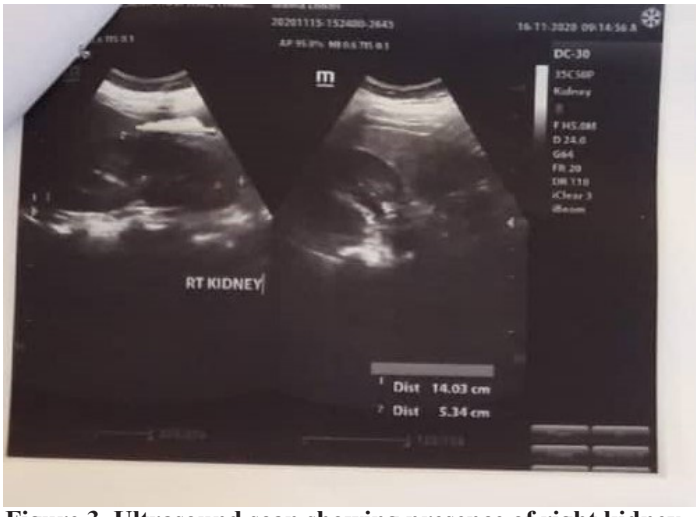

Figure 3. Ultrasound scan showing presence of right kidney only

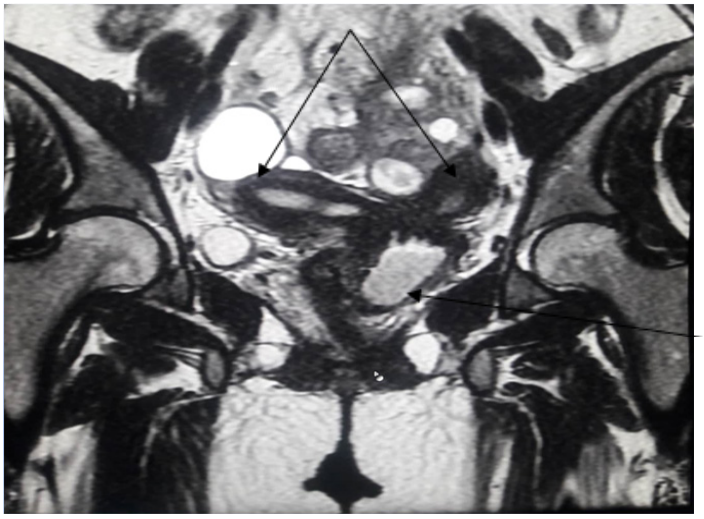

Figure 4. Coronal T2 weighted image of MRI scan showing two uteri and hematocolpos

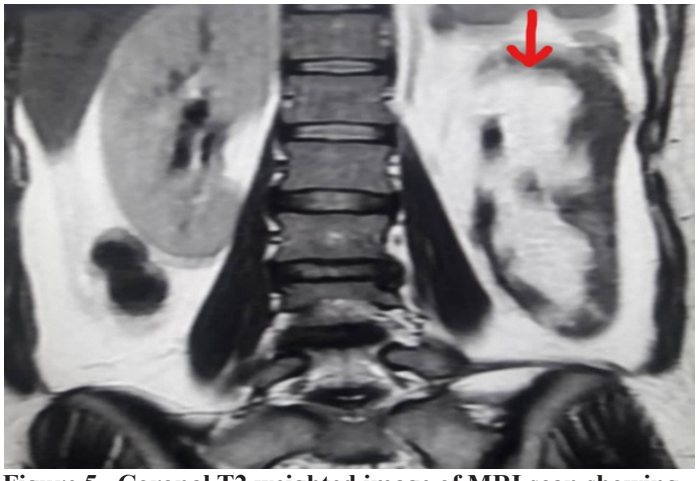

Figure 5. Coronal T2 weighted image of MRI scan showing absent left kidney

Hospital in Thimphu. She has been married for the past 18 years. Her menstrual cycles were normal but she had never conceived and received no treatment for infertility. There was no history of dyspareunia or dysmenorrhoea. On pelvic examination, the right vagina was patent and the left vaginal canal opened to left fornix of right hemivagina which was confirmed by passing uterine sound.

She had a history of being operated at a Regional Referral Hospital in the month of June 2020 for an abscess in the abdomen by a Gynaecologist. The biopsy from the abscess wall showed only inflammatory cells and no malignancy.

Axial view of contrast enhanced CT image of the abdomen showed absence of left kidney with the renal fossa occupied by the bowel loops (Figure 2). The ultrasound scan showed the presence of right kidney only (Figure 3) and coronal T2 weighted MRI scan (Figure 4 and Figure 5)) at the National Referral Hospital showed absence of left kidney, double uterus and cervix with hematocolpos on the left side of the hemivagina.

A clinical diagnosis of endometriosis was made for the patient as women with congenital uterine abnormalities with pelvic pain suffer from endometriosis[4,5] The patient was treated with injection Leuprolide Acetate 3.75mg monthly, for three months. Patient was followed up for eight months and her earlier symptom of lower abdomen pain improved. The patient had regular menstrual flow from the small communication between the two hemivagina.

DISCUSSIONS

In 1922, a case of a female patient with hematocolpos, hematometra and hematosalpinx was reported and published[6]. Nevertheless, the triad was coined only in 1971 by Herlyn and Werner when an association of renal agenesis with obstructed hemivagina was made. The component of a didelphys uterus was added by Wunderlich in 1976[1]. The syndrome is also called OHVIRA - obstructed hemivagina and renal agenesis.

The syndrome is usually diagnosed in the teenage years or soon after menarche, presenting with abdominal pain as flow of menstrual blood is blocked due to the obstructed hemivagina. When there is incomplete obstruction of hemivagina, the presentation could be at a later age. Late diagnosis could lead to complications like endometriosis and infertility[7].

The uterus, fallopian tube, cervix and upper part of vagina develop from the paired paramesonephric ducts. Uterus didelphys or uterus arcuate is formed depending on total or partial lack of fusion of the paramesonephric ducts respectively[8].

The common presentations are dysmenorrhea, pelvic and abdominal pain due to hemotocolpos as a result of obstructed hemivagina[9]. Symptoms depends on the class of the syndrome. Dysmenorrhea, intermittent mucopurent discharge and irregular vaginal bleeding is less frequent in class 1 than in class 2 [2] . Women may prevent with complications like infertility, endometriosis, and spontaneous abortion[10].

Ultrasound is a simple tool to screen this syndrome which will show absence or agenesis of one kidney. MRI scan will provide additional information – can ascertain the shape of the intrauterine cavity and the septum and assess for the presence of features of endometriosis, pelvic inflammation and adhesions[11].

Absence of one kidney by ultrasound in small hospitals should alert clinicians about this syndrome. Radiological review by Khaladkar et.al[12] showed an absence of left kidney and a study by Nishu et.al[13] found non-visualization of ipsilateral kidney by ultrasound similar to the ultrasound findings in our patient. Similar finding of an absent right kidney and possible uterine anomaly on ultrasound which was later confirmed by MRI was described by Del Vescovo et.al[11]. Therefore, ultrasound is a very useful tool for the initial diagnosis of the syndrome.

Ultrasound and MRI are the investigations of choice for diagnosis and surgical planning of HWWS. While ultrasound can be used to diagnose this condition, MRI is required for characterizing the didelphic uterus, obstructed hemivagina, and ipsilateral renal agenesis[14].

Delays in diagnosis have been attributed to the lack of understanding of this condition by radiologists, gynecologists, and pediatricians[14]

The syndrome remains unrecognized at first in the unobstructed hemivagina as there is regular flow of the menstruation. Hematocolpos is suspected only months after menarche and the diagnosis is generally made only if this syndrome is suspected[15] . This finding is similar to our case who also had regular menstruation which is why the diagnosis was probably not suspected. The didelphys uterus was not seen at laparotomy at regional hospital due to adhesions of the of bowel to each other and the fundus of uterus. Post-surgery CT revealed absence kidney and double uterus.

In a case series review of 87 patients by Fedele et.al at the University of Milan, the uterus was didelphys in 77% of cases and bicornuate in 11.5% of cases. Obstructed hemivagina was present in 95.4% of patients and ipsilateral renal agenesis was diagnosed in 95.4% of patients[16]. Zhu et al. suggested a new classification based on the presence of a completely or incompletely obstructed vaginal septum[2]. Class 1 is with complete obstruction and class 2 with incomplete obstruction of vagina with subclasses[17] (Table 1). The clinical presentation and outcomes depend on the types. The prognosis of HWW syndrome is good with early diagnosis and early treatment, except for patients with subclass 1.2 where there is cervicovaginal atresia without communicating uteri.

Our patient had a communication between two vaginas, so it was HWWS subclass 2.1. Therefore, the patient had no obstruction in the flow of menstrual blood. This could have delayed the diagnosis and missed the opportunity of corrective surgery for conception. Patients with prolonged infertility with septate uterus would benefit from corrective surgery.

We followed up our patient for six months and she is symptom free. Had the diagnosis of the syndrome been made earlier, corrective surgery could have been done. After excision of the vaginal septum and marsupialization, the pregnancy rate was 87% and 77% of them had live births[18].

CONCLUSIONS

The non obstructive type of HWWS with nonspecific abdominal symptoms delays the diagnosis and misses the opportunity to manage infertility. Gynecologists, Radiologists and General Practitioners should have high index of clinical suspicion of the syndrome for any woman presenting with primary infertility. Ultrasound findings of renal agenesis or renal abnormalities should alert the clinicians to suspect this rare syndrome. Early diagnosis and treatment are beneficial to prevent complications like primary infertility and endometriosis. In cases of primary infertility, ultrasound should be done to look for uterine anomaly and if present, HWWS should be suspected.

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