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A rare case of pelvic pain caused by Herlyn-Werner-Wunderlich Syndrome in an adult: A case report

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ABSTRACT

INTRODUCTION: Herlyn-Werner-Wunderlich Syndrome (HWWs) is a rare congenital anomaly. This abnormality has 5% percentage of the total Mullerian dysgenesis. The symptomatic patients come with varies condition, such as urinary incontinence, urinary retention endometriosis, pelvic infection and acute pelvic pain. Here we present investigation and management of HWWs with pelvic pain in adult woman.
PRESENTATION OF CASE: A 23 years old woman came with pelvic pain. We found abnormalities of HWW syndrome i.e. uterus didelphys with obstructed hemivagina and right renal agenesis through ultrasound imaging, CT scan and MRI. After concluding the diagnosis, the patient underwent vaginal septum excision and vaginoplasty procedures to relieve obstruction which was the cause of pelvic pain. The symptom was improved after surgery and she felt no more pain after a year of follow up.
DISCUSSION: The main symptom of HWWs is dysmenorrhea associated with obstruction mass or endometriosis. MRI with capability of tissue differentiation is the gold standard for diagnostic. The primary purpose of surgery is to release the obstruction and to prevent further complication.
CONCLUSION: Rarely, HWWs causes an acute pelvic pain with secondary cystic mass finding. Surgery aimed to release obstruction will improve this symptom.

1. Introduction

HWW syndrome is classified as Class III Mullerian dysgenesis, which involves uterus didelphys, obstructed hemivagina and unilateral kidney agenesis This abnormality has 5% percentage of the total Mullerian dysgenesis [1,2]. The prevalence of Mullerian dysgenesis is unclear, because many of them are asymptomatic. Based on some researches the prevalence varies from 0.4%–6.7 % [1]. Several cases have been reported to be associated with urinary incontinence, urinary retention, endometriosis, pelvic infection and acute pelvic pain [3–5]. In this case, we presented the investigation and management of HWWs which was the cause of a rare pelvic pain in adult woman. We reported and checked this case based on SCARE guidelines [6].

2. Presentation of case

A 23-years-old woman came to the clinic with complaint of menstrual pain. The patient is unmarried and has never had sexual intercourse before, she has normal activity and works at a state company every day. She has no history of smoking or other metabolic diseases. She reported a normal menstruation since it started at 12 years old. There had been no menstrual pain until she felt it in the last one year with VAS 3–4 each time of menstruation. However, the pain was getting worse lately, so she went to hospital for consultation. There were no family members who had similar complaint.

Physical examination found no significant abnormalities. Secondary sex growth was normal for her age. External gynaecology examination was also within normal limits. Further internal genital examination through the rectum found a cystic mass with a smooth surface in the right lateral region of the midline with proximal border and its origin was difficult to be determined. The uterus was slightly enlarged and there was no solid mass palpable around the cervix. Moreover, ultrasound examination was performed, exhibiting two anteflexed uteri with homogenous myometrium that were fused at the precede to the cervix. Further evaluation showed dilatation of vagina consisting of smooth, mobile hypoechoic mass; the vaginal septum lied from anterior to posterior at the middle of the vagina (Fig. 1). Both ovaries were normal; each was connected to a fallopian tube. It was difficult to define the cervix-whether single or double- while amount of blood was present in the vagina, corresponding to haematocolpos. We found left kidney in normal size, however there was no right kidney. From these findings, we concluded that the presence of uterus didelphys with obstructed right hemivagina was consistent with our suspicion of HWWs. Through ultrasound, we had not been able to assess other kidney-

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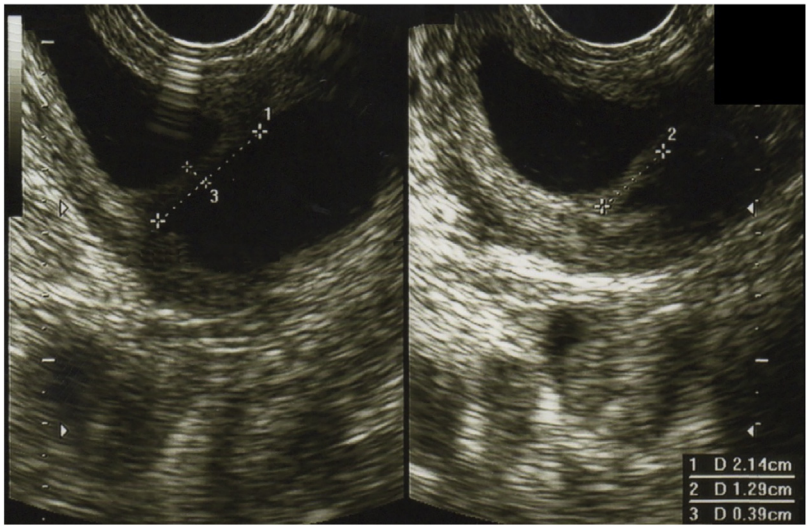

Fig. 1. Vaginal septum size 2.14 × 1.29 × 0.39 cm obstructing distal part of vagina.

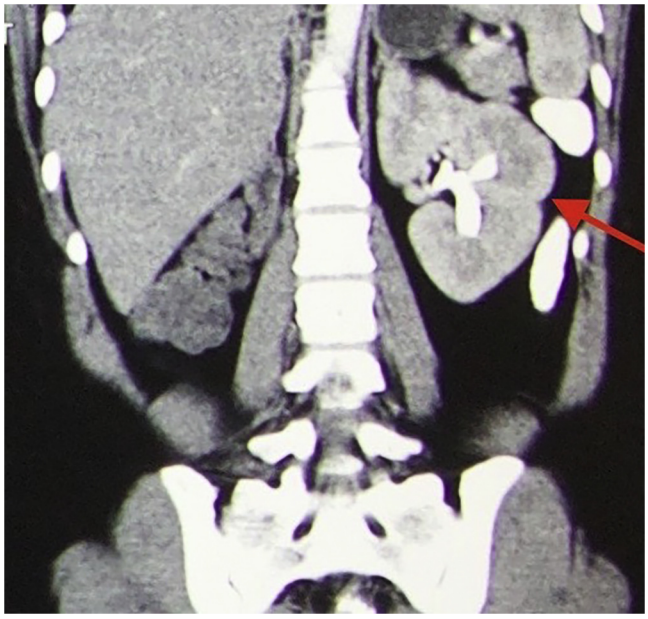

Fig. 2. Coronal section of CT scan revealed normal structure of left kidney and agenesis of right kidney.

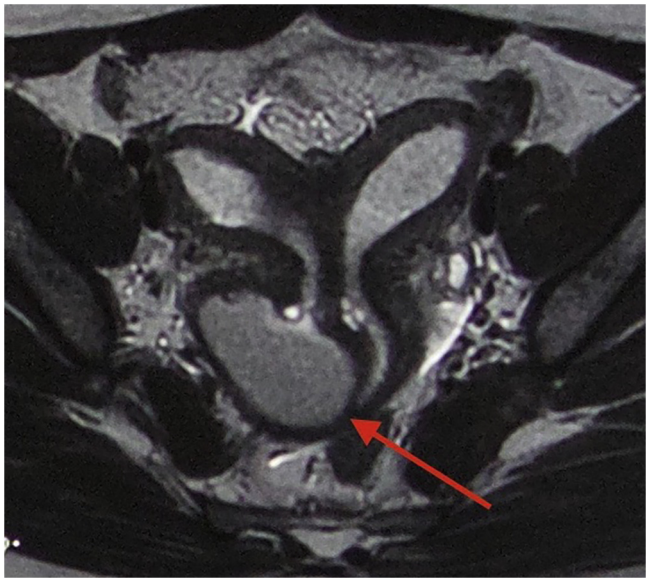

Fig. 3. MRI result revealed two normal uterine corpuses with normal endometrium and normal cervix. The distal part of vagina seen obstructed with right cystic mass (haematocolpos) appearance.

related abnormalities as well as the cervix and its association with the obstruction in the distal area of vagina.

Next, as the gold standard of tissue examination, we planned Magnetic Resonance Imaging (MRI) examination for the abdominal and pelvic areas. However, due to MRI limitation in our hospital and insurance issues, we only did one MRI examination for the pelvic by sending it to another affiliated hospital. As for assessing the abdomen, we did a Computerized Tomography (CT) scan examination in our facility. The results of CT scan showed the right renal agenesis. The left kidney had normal structure and normal ureter (Fig. 2). We performed MRI scan, which has demonstrated its importance in the diagnosis of HWW syndrome. Two complete sets of uteruses were found, each with its own corpus and cervix. The results also showed a cystic mass connected with the uterine cavity and cervical canal, obstructing the distal part of the vagina (Fig. 3). Finally, this finding highlights the accuracy of MRI scan in diagnosing HWWs

We prepared the vaginal septum excision to release the obstruction causing the pelvic pain, with informed consent about hymen damage during the procedure. The perioperative examination was unremarkable and spinal anaesthesia was used during the surgery. We performed gynaecology examination after urinary catheter insertion and found a bulging cystic mass with smooth surface from the right lateral vagina with unclear upper border (Fig. 4). A needle puncture was performed through the mass and shown blood (haematocolpos due to distal obstruction of the vaginal septum). Furthermore, 1 cm incision in the puncture area was made and widened to 3 cm. We evacuated 500 cc of blood clot. Vaginal septum with 5 mm thickness was found and excised with a diameter of 3 cm followed by vaginal reconstruction (Fig.5). There was no complication during surgery. After 3 days, the patient was in good condition and had been discharged. In the next follow-up, the patient did not complain any menstrual pain, but there was an increasing amount of menstrual blood. In one year follow up, she