CASE REPORTS

HERLYN-WERNER-WUNDERLICH SYNDROME-ROLE OF IMAGING IN TIMELY AND CORRECT DIAGNOSIS

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ABSTRACT

We present this case of uterus didelphys with right sided haematometrocolpos due to obstructed hemivagina and ipsilateral renal agenesis-Herlyn Werner Wunderlich syndrome. The condition was missed on ultrasound and diagnosed on plain CT scan abdomen and pelvis. This syndrome should be kept in mind while dealing with a young female complaining of lower abdominal pain and a pelvic mass having unilateral renal agenesis and an ipsilateral pelvic cystic lesion on ultrasound or other imaging modality. Timely diagnosis is important to avoid complications like pyocolpos and retrograde spilling of blood with consequent endometriosis. Imaging plays a major role in a correct and timely diagnosis.

Keywords: Herlyn Werner Wunderlich syndrome, Haematometrocolpos, Müllerian duct anomalies, Ohvira, Renal agenesis, Uterus didelphys.

INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome is an uncommon developmental abnormality marked by a combination of Müllerian duct and mesonephric duct abnormalities[1]. This condition is also called obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) and was first described in detail in 2006[2]. The syndrome is characterized by a triad of uterus didelphys, hemivaginal obstruction and ipsilateral renal agenesis though duplication, crossed fused ectopia or renal dysplasia may also be seen[3]. Clinically the condition usually presents with pelvic or lower abdominal pain after menarche due to haematometrocolpos because of the obstructed vagina. Examination may show a pelvic mass. Imaging investigations have an important role in the correct and timely diagnosis. Imaging features, regardless of the modality used, include type III Mullerian duct anomaly with uterine, cervical and vaginal duplication along with a blind hemivagina leading to unilateral haematocolpos or haematometrocolpos and ipsilateral renal agenesis. Differential diagnosis includes large adnexal cyst due to either cystadenoma or endometrioma. The condition is treated by surgically resecting the septum separating the two vaginal components for relieving obstruction. Successful pregnancy has been reported later on in the obstructed uterine component[4].

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CASE REPORT

A young female, age 15 years, presented to surgeon with a history of lower abdominal pain on and off. Ultrasound pelvis by a sonologist reported an absent right kidney and a large right adnexal cyst, with fine internal echoes, pushing the normal looking uterus to the left side. The surgeon planned a laparotomy and advised a plain CT scan abdomen and pelvis to confirm findings.

Plain CT scan was carried out on a single slice machine with 7 mm slice thickness. CT scan revealed absent right kidney and mild compensatory enlargement of the left kidney (fig-1). It also showed a thick walled fusiform, cystic lesion measuring 11 cm × 7.6 cm × 7.6 cm (CC × AP × Trans). The cystic lesion was located more towards the right side and extended as low as the pelvic floor with contents showing a fluid-fluid

level. A uterus like structure was seen to the left side of this cystic area which could not be separated from it lower down. Anteriorly the cystic lesion was compressing the urinary bladder while posteriorly it was associated with the rectum having clear interfaces with both (fig-2). The

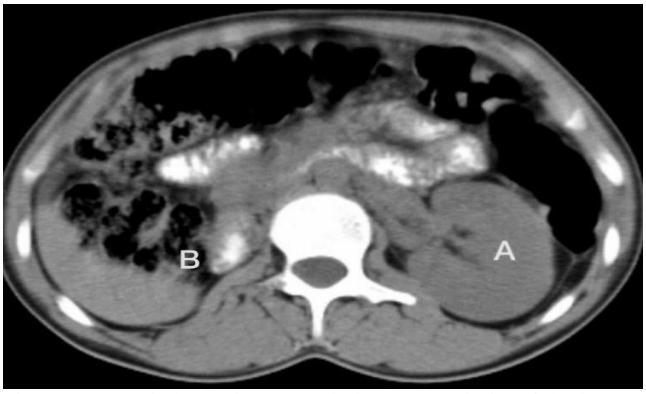

Figure-1: Axial CT image abdomen. Right sided renal agenesis. A. Left kidney with compensatory enlargement. B. Right renal agenesis.

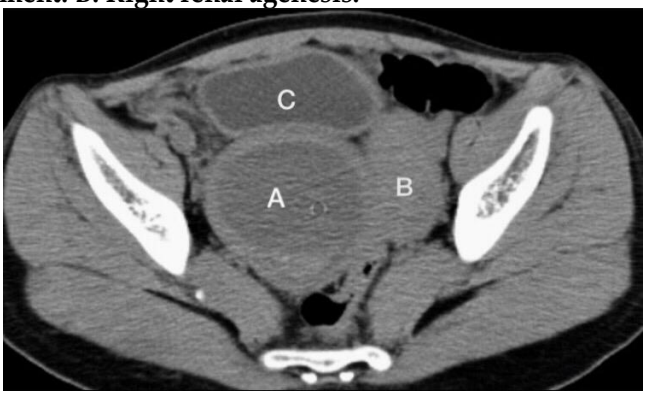

Figure-2: Axial CT image pelvis. A. Right sided haematometrocolpos with thick uniform wall and a fluid-fluid level. B. Displaced left uterine moeity. C. Urinary bladder.

presence of uterus didelphys with blind hemivagina leading to haematometrocolpos was reported along with ipsilateral renal agenesis raising the possibility of Herlyn-Werner-Wunderlich syndrome warranting MRI examination of the pelvis.

Based on CT scan report, the scheduled laparotomy for a suspected large adnexal cyst was converted into a vaginal examination under general anaesthesia. The colour of vaginal wall, the swelling and texture almost confirmed the CT findings and a wide bore needle was put though the hemivaginal obstruction followed by incision and draining all content bringing out a typical imperforate hymen blood. Following relief of obstruction, the patient was pain free and comfortable.

DISCUSSION

Herlyn-Werner-Wunderlich syndrome,with a triad of uterus didelphys, obstructed hemivagina and renal agenesis is a rare entity with the incidence of uterus didelphys ranging between 1/2,000 to 1/28,000, and in about 43% cases accompanied by ipsilateral renal agenesis[5]. Classified as a type III Müllerian duct anomaly (MDA), the condition represents about 5% of MDAs and is associated with mesonephric duct abnormalities[6]. In about half of the cases of renal agenesis there is an obstructive Müllerian duct abnormalityon the same side[3]. Right side is affected almost twice more frequently[7].

Presentation, usually at menarche, includes dysmenorrhoea and an abdominal mass due to haematometrocolpos[8]. Infection may lead to pyocolpos or pyometrocolpos presenting with fever, chills and vomiting[9]. An early diagnosis is important to avoid endometriosis as a result of retrograde tubal reflux[10]. However the diagnosis is often delayed due to the presence of regular menstruation where the symptom of cyclic dysmenorrhea is treated with medication reducing or eliminating menses.

Imaging investigations play an important role in the correct and early diagnosis. Usually the first line of imaging investigation is an abdomino-pelvic ultrasound. In experienced hands ultrasound examination can help in making a definitive diagnosis[11]. However being operator dependent, often the inexperienced, unwary sonologists are misled by the presence of a normal looking other uterine moiety and the haematometrocolpos might be mistaken for a large adnexal cyst due to the presence of a normal looking uterus displaced by the cystic mass. The sonologists, therefore, need to keep this diagnosis in mind while dealing with a case of renal agenesis and an ipsilateral pelvic cystic lesion.

Magnetic resonance imaging is the modality of choice because of its excellent soft tissue contrast and lack of exposure to ionizing radiation though some patients, as in our case, still have to undergo CT scan because of limited availability and cost of MRI[12]. MRI provides a detailed description of the pelvic anatomy because of its multiplanar imaging potential[13]. It delineates morphology of the uterus, its relationship and continuity with the obstructed and patent vaginal lumina and can show the whole abnormality very nicely including the presence of communication between the two cervices or vaginae. Besides it can also predict the nature of the fluid content and thus helps in making a better decision for the best treatment options.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

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