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Research Article

HERLYN WERNER WUNDERLICH (HWW) SYNDROME WITH HEMATOCOLPOS: A RARE CASE OF PUBESCENT FEMALE WITH PELVIC PAIN

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ABSTRACT

Herlyn Werner Wunderlich syndrome is a rare congenital mullerian anomaly consisting of uterus didelphys, hemivaginal septum and unilateral renal agenesis. Most authors reported cases of this syndrome with pre-pubertal or post pubertal onset with cyclical abdominal pain and a vaginal mass. The aim of this case report is to have a high level of clinical suspicion towards the diagnosis of this case whenever a pre-pubertal or a young adolescent girl presents with history of cyclical menstrual pain or associated features and to always investigate further by doing a ultrasonography or MRI. The high level of suspicion is, indeed the key to early diagnosis.

INTRODUCTION

Herlyn Werner Wunderlich syndrome is a rare congenital mullerian anomaly consisting of uterus didelphys, hemivaginal septum and unilateral renal agenesis. Most authors reported cases of this syndrome with pre-pubertal or post pubertal onset with cyclical abdominal pain and a vaginal mass. Our case is a 16 year female with pelvic pain which is intermittent and vague.

Case Report

A 16 yrs old female patient presented with pelvic pain which was intermittent and vague and was referred to our department for further evaluation. Ultrasonography examination showed two uterine horns and cervices with fluid collection in left sided cervix and left kidney was absent. Right kidney showed features of mild hydronephroureterosis.

Then we proceeded with MRI which showed bicornuate uterus with bicollis and a vaginal septum. Left side hemivagina and cervix showed collection of fluid within it suggestive of hematocolpos. (Fig 1) Left kidney was absent. (Fig 2) Parenchyma of right kidney was normal with mild hydronephrosis seen along with gross dilatation of ureter all along its course. (Fig 3)

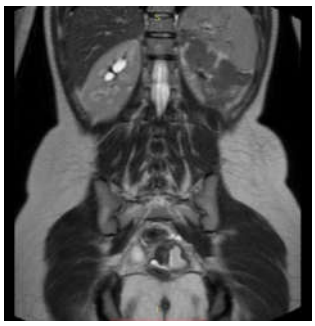

Figure 1 T2 Weighted Coronal MR Image showing left hemi – hematocolpos

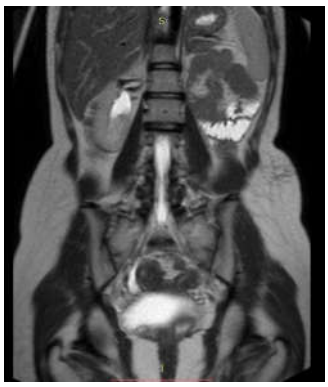

Figure 2 T2 Weighted Coronal MR Image showing absent Left Kidney with uterine didelphys

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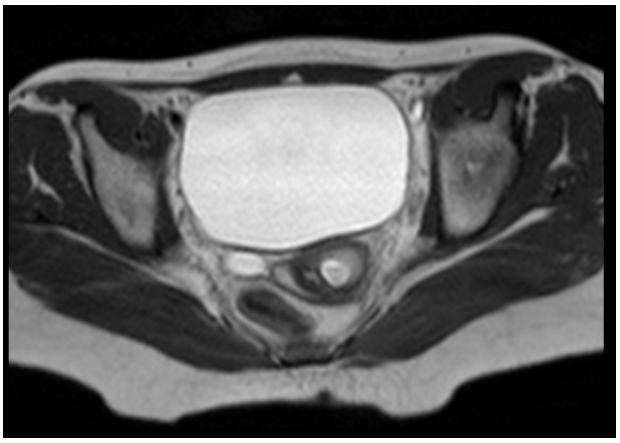

Figure 3 T2 weighted axial image showing left hemihaematocolpos with right sided hydroureter

With the above mentioned findings diagnosis of Herlyn Werner Wunderlich syndrome was made and referred to Gynecology outpatient department.

DISCUSSION

Herlyn–Werner–Wunderlich syndrome consists of unilateral renal agenesis, a blind vagina, and uterus didelphys.

The association of renal agenesis with unilateral blind hemivagina was reported as Herlyn–Werner syndrome in 1971, whereas the association of renal aplasia, bicornuate uterus with isolated hematocervix, and a simple vagina was reported by Wunderlich in 1976.[1,2] This syndrome is a rare variant within the spectrum of mullerian duct anomalies.[3] The most basic classification of mullerian ductal defects consists of

1. agenesis and hypoplasia
2. defects of vertical fusion, and
3. defects of lateral fusion

Indeed, HWWS may represent a failure of vertical and lateral fusion of mullerian structures. The actual incidence of mullerian anomalies is unknown. Patients with HWWS are usually asymptomatic until menarche when they present with ahydrometrocolpos on the side of obstructed hemivagina producing a mass effect and pain. [4]
However, the diagnosis of HWWS may be delayed by several months because menstruation is often normal. Many authors, indeed, reported cases of HWWS in post-pubertal adolescents or adult women where hematometrocolpos produces a more pronounced mass effect and pain with more complications.[5] If this syndrome is suspected, the diagnosis is simple and it can be made by ultrasound and computed tomography and/or MRI of the abdomen and pelvis.[6]

CONCLUSION

The incidence of Mullerian duct anomalies is 2-3% within the normal population. HWWS accounts for 0.1-3.5% of all Mullerian duct anomalies.

HWWS is usually diagnosed around the onset of menarche as a result of symptoms arising from the obstruction to the blood flow (haematocolpos- collection of blood in the blind / obstructed hemivagina) The diagnosis is missed on clinical examination as patients usually present with normal menstruation and non-specific abdominal pain. The radiological workup (USG and MRI) of the patient aids in diagnosis and to establish line of management. The case has been presented to help increase awareness of HWWS syndrome and the importance of radiological investigations in young females presenting with menstrual complaints. This would lead to earlier intervention and diagnosis leading to reduction in patient mortality and morbidity.

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