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CASE REPORT

HERLYN-WERNER- WUNDERLICH SYNDROME (HWW SYNDROME): A CASE REPORT

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INTRODUCTION: Congenital anomalies of the mullerian duct system can result in various urogenital anomalies and, Herlyn Werner Wunderlich syndrome (HWW syndrome) is one such rare anomaly.

This syndrome is characterized by uterus didelphys with blind hemi vagina and ipsilateral renal agenesis.[1] it is also known as OHVIRA (Obstructed Hemi vagina with Ipsilateral Renal Anomaly) syndrome.[2] Mullerian anomalies are frequently associated with renal anomalies, the incidence ranges from 0.2 to 7.4%.[3] Diagnosis of this condition is rarely made in prepubertal girls, and rather it is diagnosed after menarche. Common clinical presentation is pelvic pain shortly after menarche with an associated vaginal or pelvic mass.[4] this triad of obstructed hemi vagina, uterus didelphys and ipsilateral renal agenesis was first reported in 1950.[5]

The embryonic origin of HWW syndrome is not known, but early and accurate diagnosis of this condition is needed to enable treatment at an early stage. Surgery can provide relief from pelvic pain and preserve fertility by preventing endometriosis. Abdominal ultrasound and MRI help in the diagnosis. However, MRI has been considered as the imaging modality of choice.

CASE REPORT: A 13 years old girl presented to the department with complaints of pain in abdomen and dysmenorrhea since last 3 months. She had attained menarche 3 months back after which she had irregular bleeding with increasing dysmenorrhea and lower abdominal pain.

On examination, patient was thin built with normal height for her age. General examination showed absence of pallor. Abdomen was soft and there was no tenderness or palpable lump. Local examination was normal. Hymen was intact and her secondary sexual characters were developed as per age Tanner stage 3. MRI was done which was suggestive of uterus didelphys with left hematocolpos extending into left endocervical canal, renal agenesis and compensatory right renal hypertrophy.

Patient was admitted and examined under anesthesia with diagnostic laparoscopy was planned. Per speculum examination revealed a normal vagina with normal cervix on right side and high up in the vaginal on left side a tense bulge was seen. Per vaginal examination showed a tense bulge of approximately 4x4cm.

A transverse incision of 1.5cm was taken over the left bulge and approximately 200ml chocolate colored fluid was drained suggestive of hematocolpos. A normal cervix was visualized inside. Laparoscopy was done and findings were suggestive of two normal size uteri with band connecting the two. The two bilateral ovaries were normal as were both fallopian tubes. There were few endometriotic lesions in the pelvis. The patient withstood the surgery well and was discharged on day 3.

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DISCUSSION: HWW syndrome is a type of mullerian duct anomalies (MDA) associated with mesonephric duct mal development. MDA results due to agenesis, defective ventral or lateral fusion or resorption failure of mullerian duct.[6] this syndrome comes under class III of American Fertility Association (AFS) classification.

It is usually discovered at puberty with dysmenorrhea, increasing pelvic pain and palpable mass which is due to retained long standing menstrual flow. It can also rarely be seen in patients with primary infertility when the vaginal septum is incomplete.[7] Some patients present with complications like pyohematocolpos, pyosalpinx or peritonitis and long term complications including endometriosis, pelvic adhesion and infertility. [8]

For diagnosis, ultrasonography can be used as primary modality that can show utero vaginal duplication, hematocolpos or hematometra and absent ipsilateral kidney. However, MRI has been considered accurate imaging method for diagnosis.[9]

CONCLUSION: The diagnosis of this syndrome is difficult due to its rarity and high index of suspicion is required for its diagnosis. Early detection in teenage girls is important as immediate surgical resection of the vaginal septum obstructing the outflow tract can provide pain relief as well as prevent further complications that may lead to infertility.

Figure 1: Laparoscopic image suggestive of two uteri with a band like structure connecting the two and endometriotic patches in pouch of Douglas. Bilateral fallopian tubes and ovaries appear normal.

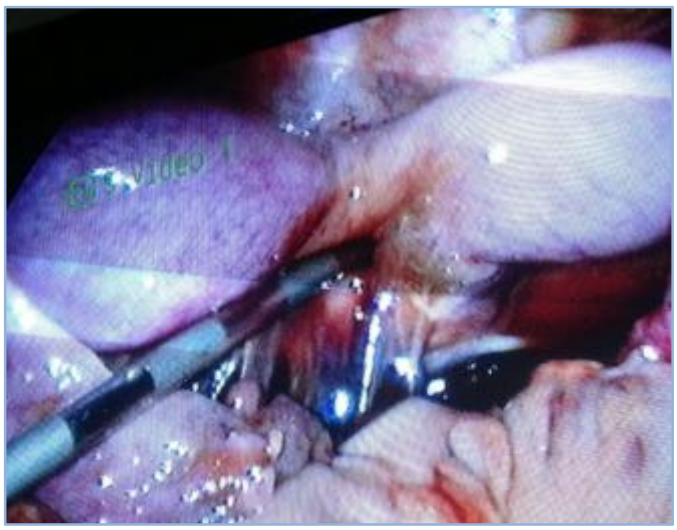

Figure 1

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