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HERLYN-WERNER-WUNDERLICH SYNDROME WITH ENDOMETRIOSIS OF OVARY AND FALLOPIAN TUBE- A RARE CASE REPORT

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ABSTRACT The Herlyn Werner Wunderlich Syndrome (HWWS) is an extremely rare type of Mullerian Duct Anomaly characterized by didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis. We present a case of HWWS in an 18 year old female with associated complication of Endometriosis of ovary and fallopian tube and discuss the role of a histopathologist in the diagnosis and management of this rare entity.

KEYWORDS : Herlyn Werner Wunderlich Syndrome, Mullerian Duct Anomalies, Endometriosis

Introduction:

Herlyn Werner Wunderlich syndrome is a rare congenital anomaly with very few cases reported till date. It is characterized by uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. It usually presents after menarche with progressive pelvic pain during menses, dysmenorrhea, pelvic mass due to hematocolpos or it can also be an accidental finding on radiological investigations. The triad was first reported in 1971 by Herlyn and Werner, and again in 1976 by Wunderlich, hence the name [1]. Associated complication of endometriosis is reported in about 1/5th of the patients and is a result of delayed clinical attention due to lack of awareness of this rare entity.[2]

Case Report:

An 18 year old unmarried female came to the hospital with the chief complaints of pain in abdomen since two weeks and dysmenorrhoea sine two months.

Patient had attained menarche at 12 years of age and had no menstrual irregularity, until two months back.

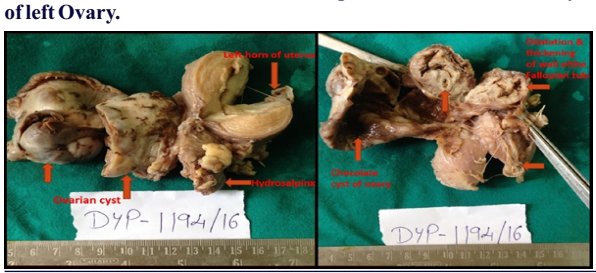
Routine investigations were normal. USG revealed absence of left kidney with compensatory hypertrophy of right kidney and a didelphys uterus with left sided hematocolpos and hematosalpinx with blind hemivagina. Left ovary showed a large cystic mass suggestive of chocolate cyst.

CT scan confirmed the USG findings. Based on these findings, a diagnosis of Herlyn Werner Wunderlich syndrome was made.

The patient was subsequently taken up for surgery and left sided Salpingo- Oopherectomy with left horn of uterus was removed along with reconstruction of the vagina.

Grossly we received part of uterus (body and fundus) measuring 5 x 4 x 3.5cm with attached left sided fallopian tube and ovary with cyst. Fallopian tube appeared dilated with thickened wall measuring 5cm in length and 3.5cm in diameter. Ovary measuring 7.5 x 6.5 x 5cm with already cut opened cyst measuring 5.5 x 5cm with thickness of 0.5cm. The fallopian tube and the cyst cavity showed brownish material.

Figure 1: Gross images showing left horn of Uterus with attached left sided dilated and thickened Fallopian tube and chocolate cyst of left Ovary.



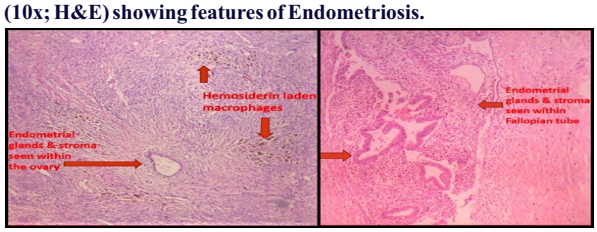
Mutliple sections were taken from the uterus, fallopian tube, normal part of ovary and ovarian cyst for microscopic examination.

Sections from fallopian tube showed foci of endometrial glands within the wall along with oedematous, thickened wall showing chronic inflammatory cell infiltrate.

Setions from ovarian cyst showed cyst wall lined by flattened cuboidal lining epithelium along with foci of endometrial glands and plenty of hemosiderin laden macrophages.

Figure 2: Microscopy

Section from Ovary (left) (10x; H&E) and Fallopian tube (right) (10x; H&E) showing features of Endometriosis.



Discussion:

Herlyn Werner Wunderlich syndrome is an example of Mullerian duct anomaly and constitutes approximately 0.1 to 3.5% of all Mullerian duct abnormalities. [3]

Patient usually present post- menarche with pelvic pain, dysmenorrhea and associated pelvic mass due to collection and stagnation of menstrual products in the blind uterine horn as was seen in our case.[1] Didelphic uterus occurs due to embryologic arrest during the 8th week of gestation which affects the Mullerian and Metanephric ducts. Exact cause of this developmental defect is not known.[4]

The Wolffian duct gives rise to ipsilateral ureteric bud which is responsible for formation of the kidney. If the Wolffian duct on one side is congenitally absent, the kidney and ureter of that side will fail to fuse at midline. On the side lacking the Wolffian duct, the Mullerian duct displaces itself laterally and cannot come in contact with the urogenital sinus in the centre resulting in a blind sac, imperforate or obstructed hemivagina. However the distal part of the vagina is not affected and develops normally.[4] HWWS is the rarest of all Mullerian duct anomalies but has the best prognosis. [4]

The primary diagnosis of HWW syndrome is made radiologically, with MRI and USG being the diagnostic modalities of choice. However, the gold standard for diagnosis is laproscopy as it has the added benefit of performing therapeutic drainage of hematocolpos, vaginal septotomy and marsupialisation (5) The role of histopathology in HWW syndrome is essential to rule out the associated complications, most common being Endometriosis.

Endometriosis refers to presence of endometrial glands and stroma in

abnormal locations outside the uterus whereas abnormal distribution of endometrial tissue within the myometrium is termed as Adenomyosis. Diagnosis of endometriosis is made microscopically in the presence of any 2 out of 3 criteria: Endometrial glands, Endometrial stroma and Hemosiderin laden macrophages.

The histogenesis of endometriosis has been a debatable matter for years. Currently the following 3 theories are described:

1) **Retrograde Menstruation** theory is based on the assumption that ectopic endometrial tissue is transplanted from the uterus to an abnormal location by way of fallopian tubes due to regurgitation of menstrual blood. This theory is the most widely accepted theory.
2) **Metaplastic theory** suggests that ectopic endometrium develops in situ from local tissues by metaplasia of the coelomic epithelium.
3) **Vascular or Lymphatic Dissemination** explains the development of endometrial tissue at extrapelvic sites by this route.

The results seen in multiple studies suggesting that endometriosis is seen in about 20% of the cases of HWWS supports the Retrograde Menstruation Theory of formation of endometriosis as there is prolonged stasis and retrograde flow of menstrual products due to hematocolpos.

Other associated complications of HWW syndrome are infections, pyocolpos, pelvic adhesions which in turn might cause obstruction of the genital organs.

About 87% of the patients of HWWS undergo successful pregnancy after surgical management, however 23% patients carry the risk of abortion.[5]

Conclusion:

Early diagnosis and prompt surgical management of HWWS and other Mullerian duct anomalies can prevent dire complications and preserve fertility in the patient. Hence awareness of HWWS and other Mullerian Duct anomalies is essential in current clinical practice.

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