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Case Report

HERLYN-WERNER-WUNDERLICH (HWW) SYNDROME AND ITS SURGICAL MANAGEMENT- A CASE REPORT

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ABSTRACT

Herlyn- Werner-Wunderlich is a rare congenital anomaly which is generally detected just after menarche. In this case report a 11year old girl presented with cyclical lower abdominal pain and complaints of hypomenorrhea. After radiological imaging it was found that she was having uterine didephys and unilateral obstruction and blind hemivagina. She was relieved of her symptoms by surgically opening the blind hemivagina and the dilatation of the strictured cervix.

KEY WORDS: Mullerian anomaly, Renal agenesis, Hematocolpos, Hematometra, Blind hemivagina

INTRODUCTION

Approximately 10% of infants are born with some abnormalities of the genitourinary system[1]. The Herlyn–Werner–Wunderlich syndrome is a rare congenital anomaly characterised by uterus didelphys with blind hemivagina and ipsilateral renal agenesis[2]. This syndrome was suspected for the first time in a young woman with regular menstruation and gradually increasing pelvic pain and a pelvic mass after menarche in 1922 and it was described in literatures. The true incidence of this anomaly is still unknown, but it has been reported between 0.1% and 3.8%[3]. Renal agenesis with an ipsilateral blind hemivagina was initially described in 1971 by Herlyn and Werner as Herlyn-Werner syndrome[4]. Wunderlich found an association of right renal aplasia with a bicornuate uterus and a single vagina in the presence of an isolated hematocervix In 1976[5]. In this case report we describe about a 11year old girl with complaints of hypomenorrea and progressive cyclical lower abdominal pain since menarche and her surgical management.

CASE REPORT

A 11-year-old girl presented in a Gynaecological OPD of a tertiary care medical college of West Bengal with cyclical lower abdominal pain during menstruation since her menarche and complaints of scanty menstruation. She had an average built, adequate nutrition, and of good intelligence quotient. Her general survey was within normal limits. On abdominal examination there was no tenderness or rebound tenderness in any quadrant. Tanner stage for breast was stage 2 and tanner stage for pubic hair was stage 3, without any axillary hair. On external vaginal examination no visible abnormality was identified.

Radiological investigations were done where ultrasonography of whole abdomen revealed she is having two separate uterine body and cervix

with two complete separate endometrial echo. Localised echogenic collection (62*29mm) seen in left sided lower endometrial cavity and cervical canal; suggestive of hematometra and haematocolpos on left side. Right endometrial cavity and cervix appear normal. Vagina could not be visualised properly. Right kidney normal and left kidney not visualised in its normal location or in any ectopic position.

MRI revealed two separate uterine horns having separate endometrium and myometrium. Complete thin septa noted in between two cervical canals. Left cervical canal is hugely distended with collection having blood products within. Right cervical canal is compressed, showing thin T2 hyperintense fluid. The two cervical canal is opening into the vagina with narrowing at cervico-vaginal junction suggesting stenosis. Left ovary is not visible; right ovary identified with tiny follicles. Left kidney is not visible. Right kidney enlarged with compensatory hypertrophy. Features are suggestive of uterine didelphys with cervicovaginal stenosis and left sided hematometra and congenital absence of left kidney.

After proper blood investigations, decision for examination and dilatation of cervix under anaesthesia was taken to relieve her symptoms. On 09/04/2021; exploration and dilatation was attempted and intra operatively it was found that only right cervical canal is visible and left cervix was not at all visualised. Right cervix was also having stenosis and after mild dilatation foul smelling blood came out; suggesting old menstrual blood. Per rectally a boggy mass was being felt even after evacuation of blood but the left cervix remained unapproached in this sitting. Post operatively her pain was reduced and she had no other complaints. Before discharge another ultrasonography was done in order to evaluate the patient on later stages. This ultrasound revealed 50*15 mm localised collection in left sided lower endometrial cavity. Right sided cervix appeared normal. But on her next menstrual cycle she again started having dysmenorrhea and scanty menstruation.

She was planned for another setting of evaluation under anaesthesia during her menstrual period. After proper asepsis and in lithotomy position, with the help of two sims speculum, one cervix is identified at right side. A vaginal bulge with bluish hue was identified on left side. Incision made over the bulge, foul smelling brownish fluid; suggesting old menstrual blood came out. Incision extended and it is identified that there is a longitudinal vaginal septum. Left sided external os identified and it is dilated with dilator. The incision line is oversewn with 3-0 vicryl to secure hemostasis. The two vaginas are intercommunicating in the lower third of the vaginal canals. The girl was discharged after 2 days with advice of local oestrogen cream application and oral oestrogens.

On follow up scans it was found that there is no stricture and no hematocolpos or hematometra is being formed. On the next menstrual cycle there was no complaints of dysmenorrhea and hypomenorrhea.

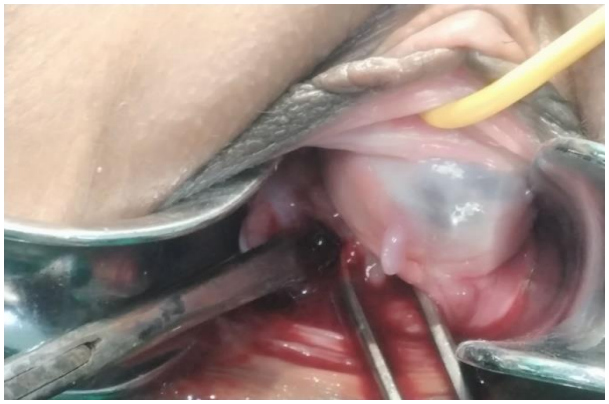


FIG 1: Intra-operative photo showing bluish hue and a bulge on left side of the vaginal canal.

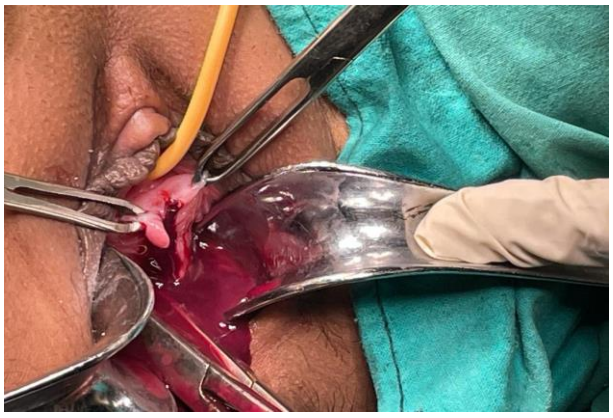


FIG 2: on giving a sharp nick at the most prominent area; evacuation of altered blood noted.

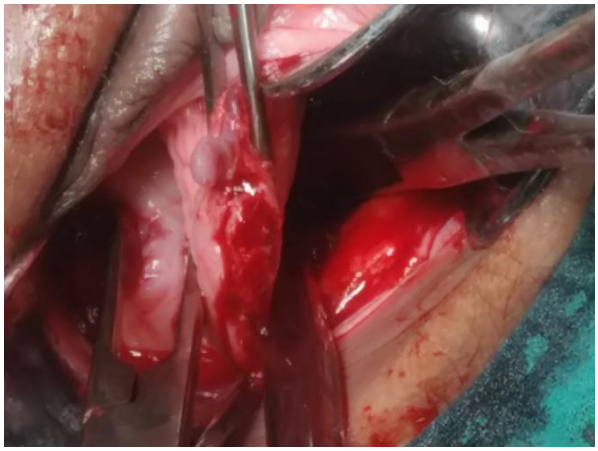


FIG 3: opening of the blind vaginal canal

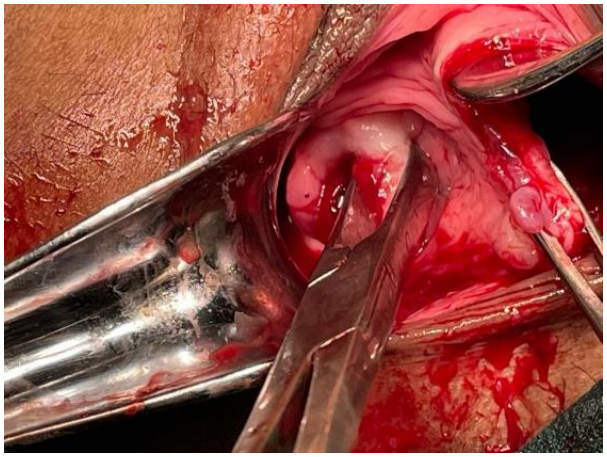
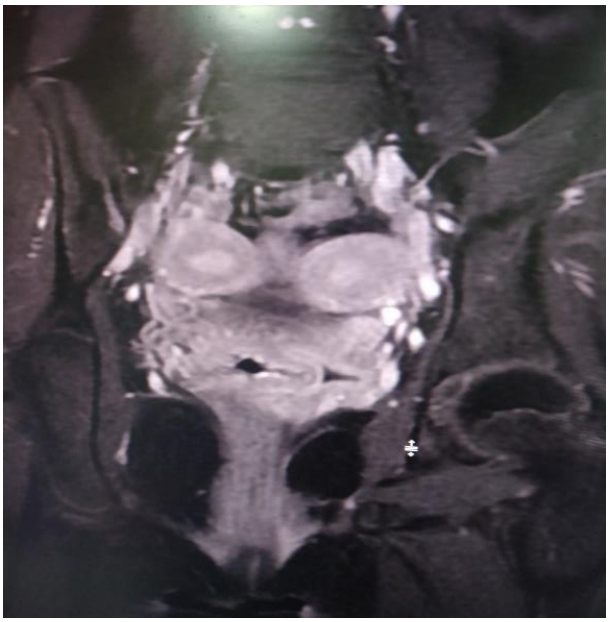


FIG 4: Image showing healthy right cervix and the newly created left hemivagina.



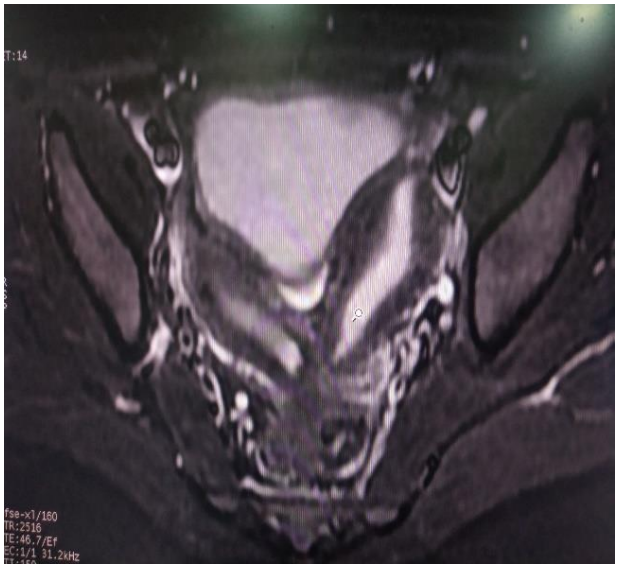


FIG 5: Post operative MRI showing didelphys uterus without any gross distension.

DISCUSSION

The urogenital system is derived from Intermediate mesoderm and the primitive urogenital sinus; which is a part of the cloaca. The uterine tubes are derivative of Paramesonephric duct; mesodermal origin. The uterus is formed by fusion of right and left paramesonephric ducts (uterovaginal canal). As the thickness of the myometrium increases, the unfused horizontal parts of the two paramesonephric ducts come. External genitalia formed from swellings that appear around the urogenital membranes. The two paramesonephric ducts get fused and give rise to uterine body and cervix. Vagina on the other hand has double origin; upper one third is derived from uterine canal and lower two thirds from urogenital sinus.

Lack of fusion of paramesonephric ducts in a localised area or throughout the length of the ducts may explain all possible uterine congenital anomalies. Our case comes under the classification where there is complete obstruction of hemivagina; uterus behind the septum is completely isolated from the contralateral uterus. There is no communication between the two uterus and vagina. Hematocolpos might occur after few months of menarche[6].

FIG 6: Schematic diagram showing complete obstruction of a hemivagina with isolated uterus and formation of hematocolpos and hematometra. [according to the new classification system of Herlyn Werner Wunderlich syndrome]

Wolfian Duct is responsible for giving rise to ipsilateral ureter and kidney. On failure of formation of Wolfian duct (Left side for the present case scenario), the ipsilateral Mullerian duct gets displaced laterally and a blind sac and hemivagina is created from the Mullerian duct system; which fails to fuse with the other side and gives rise to a blind uterine sac leading to formation of hematocolpos and hematometra after menarche. The distal portion of vagina remains unaffected as it has a different origin[7].

CONCLUSIONS

The Herlyn-Werner-Wunderlich (HWW) syndrome being extremely rare there needs to be a high amount of suspicion and clinical eye to isolate it from other mullerian anomalies. It is very commonly mistaken with other mullerian anomalies and the complaints of hypomenorrhea and dysmenorrhea are not taken into consideration. The diagnosis done via ultrasonography and MRI is of utmost help as it is noninvasive and do not cause any additional radiation.

BIBLIOGRAPHY

1. Berek, J.S. Anatomy and Embryology. In: Berek, J.S, Berek, D.L (eds.) Berek & Novak's Gynecology. NEW DELHI: Wolters Kluwer (India); c2020. p. 49.
2. Aveiro AC, Miranda V, Cabral AJ, Nunes S, Paulo F, Freitas C. Herlyn-Werner-Wunderlich syndrome: a rare cause of pelvic pain in adolescent girls. BMJ Case Rep. 2011 Jul 15;2011:bcr0420114147.
3. Zurawin RK, Dietrich JE, Heard MJ, Edwards CL. Didelphic uterus and obstructed hemivagina with renal agenesis: case report and review of the literature. J Pediatr Adolesc Gynecol. 2004 Apr;17(2):137-41.
4. Jia, G., Chai, W., Cui, M., Wen, Y., Cui, L., & Gong, F. (2018). A case report on Herlyn-Werner-Wunderlich syndrome with spontaneous abortion. Medicine, 97(36), e12004.
5. Wunderlich M. [Unusual form of genital malformation with aplasia of the right kidney]. Zentralbl Gynakol. 1976;98(9):559-62.
6. Zhu L, Chen N, Tong JL, Wang W, Zhang L, Lang JH. New classification of Herlyn-Werner-Wunderlich syndrome. Chin Med J (Engl). 2015;128(2):222-225. doi:10.4103/0366-6999.
7. Fedele L, Motta F, Frontino G, Restelli E, Bianchi S. Double uterus with obstructed hemivagina and ipsilateral renal agenesis: pelvic anatomic variants in 87 cases. Hum Reprod. 2013 Jun;28(6):1580-3.

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