A Case of Herlyn-Werner-Wunderlich Syndrome (Uterus Didelphys, Blind Hemivagina and Ipsilateral Renal Agenesis) and Coexistence of a Vertebral Anomaly

Herlyn-Werner-Wunderlich Sendromuna (Uterus Didelfiz, Kör Hemivajina ve İpsilateral Renal Agenezi) Eşlik Eden Vertebral Anomali Olgusu

Nazire ÖZTÜRK,[a]
Uğraş ŞAHİN,[a]
Uğurkan ERKAYIRAN,[a]
A. Sertaç BATIOĞLU[b]

[a]Clinic of Gynecology and Obstetrics, Ankara Education and Research Hospital,
[b]Clinic of Reproductive Endocrinology, Dr. Zekai Tahir Burak Women's Health, Research and Education Hospital, Ankara

Geliş Tarihi/*Received:* 14.02.2011
Kabul Tarihi/*Accepted:* 29.04.2011

Yazışma Adresi/*Correspondence:*
A. Sertaç BATIOĞLU
Dr. Zekai Tahir Burak Women's Disease, Education and Research Hospital, Clinic of Reproductive Endocrinology, Ankara,
TÜRKİYE/TURKEY
asbatioğlu@gmail.com

ABSTRACT A 17-year old woman was admitted to our department with recurrent pelvic pain at the time of menses. The patient's menarche had been at the age of 14 years, thereafter, she had menstruated regularly at monthly intervals with scanty menstrual flow and severe dysmenorrhea. An abdominal ultrasound scan showed two widely divergent uterine horns with no communication between them. There was hematometra on right hemiuterus. Intravenous pyelography didn't demonstrate right kidney. All of these findings revealed the triad of obstructed hemivagina and uterus didelphys as well as an ipsilateral renal anomaly, (Herlyn-Werner-Wunderlich Syndrome) Besides these anomalies, there was also scoliosis. Strasman metroplasty was performed. In conclusion, in the case of severe primer dysmenorrhea seen shortly after menarche, Herlyn-Werner-Wunderlich Syndrome must be investigated in differential diagnosis.

Key Words: Uterus; vagina; scoliosis

ÖZET 17 yaşında hasta, adet dönemlerinde tekrarlayan pelvik ağrı ile kliniğe başvurdu. Hastanın ilk adet yaşı 14 olup, her ay düzenli olarak miktarı az ama şiddetli ağrılı adet görüyordu. Abdominal ultrason incelemede aralarında bağlantı görülmeyen birbirinden uzaklaşan iki geniş uterus bulundu. Sağ hemiuterusda hematometra vardı. İntravenöz pyelografide sağ böbrek görüntülenemedi. Bu bulgular, kör hemivajina ve uterin didelfiz yanında aynı taraf böbrek anomali triadını gösterdi (Herlyn-Werner-Wunderlich sendromu). Bu anomaliler yanında hastada ayrıca skolyozis tespit edildi. Aile ile birlikte karar verilerek Strassman operasyonu uygulandı. Sonuç olarak, adetten kısa süre sonra görülen şiddetli primer dismenore olgularında Herlyn-Werner-Wunderlich sendromunun ayırıcı tanıda araştırılmasının gerekeceği vurgulandı.

Anahtar Kelimeler: Uterus; vajina; skolyoz

Turkiye Klinikleri J Gynecol Obst 2012;22(3):188-90

The combination of obstructed hemivagina and uterus didelphys was first reported in 1922,[1] however the triad of obstructed hemivagina and uterus didelphys as well as an ipsilateral renal anomaly was not reported until 1950.[2] The triad called as Herlyn-Werner-Wunderlich Syndrome since 1983.[3] In the general population the reported incidence for this anomaly is 0.1-3.8%.[4]

The most common clinical presentation is the onset of pelvic pain and/or dysmenorrhea shortly after menarche, in association with the finding of a vaginal/pelvic mass. A patient with dysmenorrhea from a double uterus and obstructed hemivagina is a diagnostic dilemma because the

menses are regular. Sometimes an accompanying anomaly can be seen with the syndrome, Herein we represent a case of Herlyn-Werner-Wunderlich Syndrome and coexistence of a vertebral anomaly (scoliosis).

CASE REPORT

A 17-year old woman was admitted to our department with recurrent pelvic pain at the time of menses. The patient's menarche had been at the age of 14 years, thereafter, she had menstruated regularly at monthly intervals with scanty menstrual flow and severe dysmenorrhea.

An abdominal ultrasound scan showed two widely divergent uterine horns with no communication between them. The thicknesss of endometrium of left hemiuterus was 7 mm and there was hematometra on right hemiuterus (Figure 1). Her bilateral ovaries were normal. Intravenous pyelography didn't demonstrate right kidney, there was compensator hypertrophy on left kidney. Besides these anomalies, there was also scoliosis (Figure 2).

With the informed consent of patient and her family, uterus unification was made with performing strassman metroplasty. The suture which was made on medial surfaces of both uterus elongated to enclose cervix and distal 1/3 of both hemivagina (The left one was obstructed). The semilunar surfaces of both hemiuterus has been suturated from anterior to posterior (Figure 3). The patient was asymptomatic after surgery. Also office hysteroscopy was made to confirme communication between both hemivagina and hemiuterus two months later.

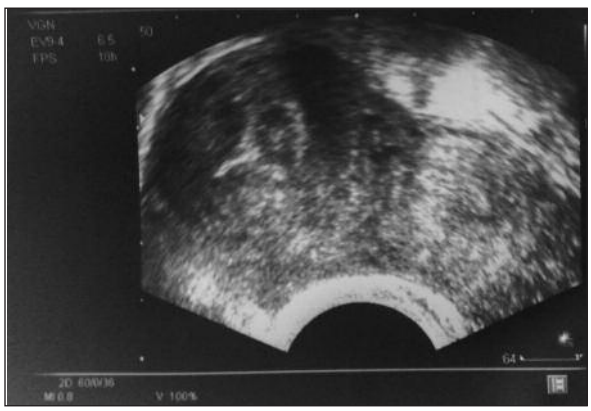

FIGURE 1: Hematometra on right hemiuterus.

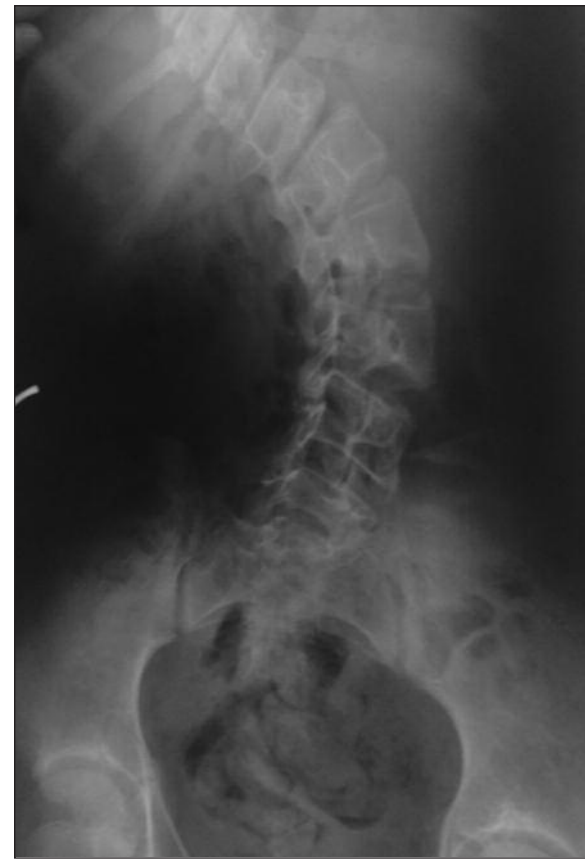

FIGURE 2: Vertebral anomaly.

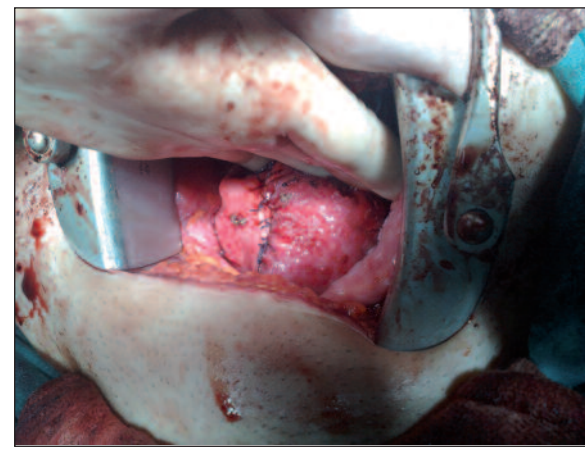

FIGURE 3: Uterus unification made with strassman metroplasty.
(See for colored form http://jinekoloji.turkiyeklinikleri.com/)

DISCUSSION

Mullerian duct anomalies are congenital anomalies of the female genital tract that result from nonde-

of the female genital tract that result from nondevelopment or nonfusion of the mullerian ducts on failed resorption of the uterine septum.[4-6] The etiologic factors are unknown, but may include an insult during the first trimester or polygenic/multifactorial inheritance.

The complete form of didelphys is characterized by 2 hemiuterus, 2 endocervical canals with cervices fused at the lower uterine segment. Each hemiuterine is associated with one fallopian tube. The association of uterus didelphys with obstructed hemivagina with renal agenesis ipsilateral to the side of obstruction can be explained by embryologic arrest at the 8th gestation week, which simultaneously affects the mullerian and metanephric ducts.

Early and accurate diagnosis is vital because untreated patients may develop retrograde tubal reflux and endometriosis. It may cause infertility and obstetric complications later in life.

This syndrome can be easily overlooked, because patients with this syndrome menstruate normally.

Imaging modalities to diagnose this condition include ultrasonography, magnetic resonance imaging and three dimensional computed tomography angiography and laparoscopy.

CONCLUSION

There are a lot of types of mullerian duct anomalies. Some of these are easily diagnosed but for others to diagnose a high index of suspension is necessary.

A greater awareness of the syndrome of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis should lead to its prompt diagnosis, allowing for early and appropiate sur-

REFERENCES

1. Purslow CE. A case of unilateral heamatocolpos,hematometra and haematosalpinx. J Obstet Gynaecol Br Emp 1922;29(1): 643.
2. Embrey MP. A case of uterus didelphys with unilateral gynatresia. Br Med J 1950;1(4657): 820-1.
3. Karag"ozov I. [Herlyn-Werner-Wunderlich syndrome]. Akush Ginekol (Sofiia) 1983;22(1):70-6.
4. 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;17(2):137-41.
5. Madureira AJ, Mariz CM, Bernardes JC, Ramos IM. Case 94: Uterus didelphys with obstructing hemivaginal septum and ipsilateral renal agenesis. Radiology 2006;239(2):602-6.
6. Polat İ. [Herlyn-Wener-Wunderlich syndrome: didelphic uterusand obstructed hemivagina with ipsilateral renal agenesis: a case series]. Turkiye Klinikleri J Gynecol Obst 2008;18(4): 284-8.