Olgu sunumu / Case report

UTERUS DIDELFIS, OBSTRÜKTE HEMIVAJEN VE İPSILATERAL RENAL AGENEZI: OHRIVA SENDROMU

Uterus Didelphys, obstructed hemivagina and ipsilateral renal agenesis: OHRIVA Syndrome

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Özet
Herlyn-Werner-Wunderlich Sendromu uterus didelfis, obstrükte hemivajen ve ipsilateral renal agenezi ile karakterize nadir görülen bir anomalidir. Bu sendroma sahip hastalar sıklıkla menarştan kısa süre sonra başlayan dismenore, siklik abdominal ağrı, akut abdomen ve abdominal kitleden yakınırlar. Bu olgu sunumunda 13 yaşında Herlyn-Werner Wunderlich Sendromlu bir vaka sunmayı amaçladık
Anahtar kelimeler: mülleryan anomali, obsrükte hemivajen, OHRIVA sendromu, uterus didelfis, Herlyn- Werner Wunderlich Sendromu

Abstract
Herlyn-Werner-Wunderlich syndrome is a rare urogenital anomaly characterized by uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. Patients with this syndrome frequently complain about dysmenorrhea, cyclic abdominal pain, acute abdomen and abdominal mass soon after menarche. İn this case report, we aimed to report a thirteen-year old girl who had Herlyn- Werner Wunderlich Syndrome.
Key Words: mullerian anomaly, obstructed hemivagina, OHRIVA syndrome, uterus didelphys, Herlyn- Werner Wunderlich Syndrome

Geliş Tarihi / Received: 13.10.2013, Kabul Tarihi / Accepted: 23.11.2013

Introduction

Uterus didelphys together with obstructed hemivagina is seen rarely. Although patients with uterus didelphys and obstructed hemivagina are usually asymptomatic until menarche, dysmenorrhea becomes the major complaint after menarche. İn this case report, we aim to present a patient having complaint of dysmenorrhea and diagnosis of Herlyn- Werner Wunderlich Syndrome, characterized by uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis.

Case

Thirteen years old girl admitted to our clinic with a chief complaint of dysmenorrhea. Her menarche was 4 months ago and her menstruation periods were regular. Her medical history was unremarkable. In physical examination, tenderness and 4x4 cm mass were observed in right lower quadrant of abdomen. Rebound and defense were not seen. Bulging in right vaginal wall was observed in pelvic examination with virgin speculum. Rectal examination revealed a cystic mass on the right side of vagina. Pelvic ultrasonography (USG) showed 35x47 mm cystic lesion on the right inferolateral side of uterus. To clarify diagnosis magnetic resonance imaging (MRI) were performed. MRI revealed uterus didelphys and cystic lesion between right cervix and left proximal vagina which were thought hydro-hematocolpos (figure-2a, 2b, 2c, 2d). Intravenous pyelography (IVP) could only visualize the left urinary system (figure-1). Coexistence of uterus didelphys, unilateral obstructed hemivajina, hidro-hematometrocolpos and right renal agenesis suggested Herlyn-Werner-Wunderlich syndrome

for diagnosis. Operation was planned and right vaginal wall was insized from its most bulging section. 50 cc hemorrhagic fluid were drained. Insicion were expanded and vaginal septum was resected completely to combine both hemivagina.

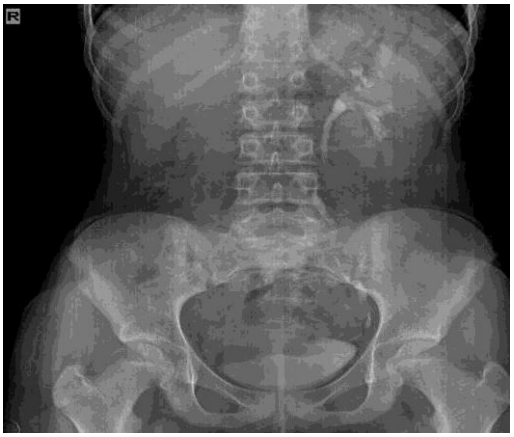

Figure-1

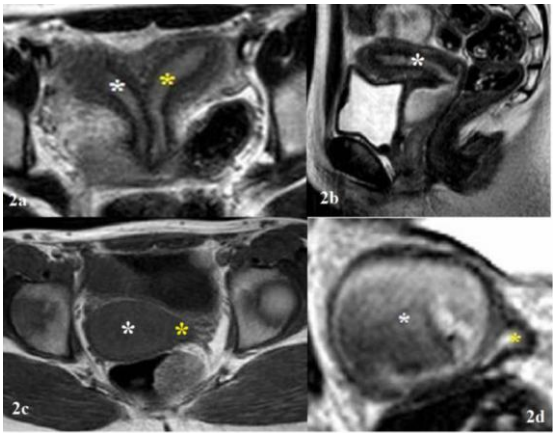

Figure-2a, 2b, 2c, 2d

Discussion

Herlyn-Werner-Wunderlich syndrome, coexistence of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis, is a rare urogenital anomaly(1). This syndrome was also called as OHVIRA (Uterus didelphys with Obstructed Hemivagina and Ipsilateral Renal Agenesis) syndrome. Although the exact etiopathogenesis is not known, it is suggested that incomplete fusion of mullarian duct might cause uterus and cervix duplication. Also fusion defect between mullerian duct and urogenital sinus might cause obstructed hemivagina(2). Diagnosis can be done usually short after menarche with beginning of complaints(3). Generally, patients complain about dysmenorrhea, abdominal mass, cyclic abdominal pain and acute abdominal pain(4). Differential diagnosis should be made for ovarian torsion and acute appendicitis. Attention to patints` history and physical examination is necessary not to skip diagnosis, because dysmenorrhea is common complaint in this age group and patient can menstruate regularly from other side of vagina. Early diagnosis not only resolves symptoms but also prevents endometriosis formation later(5). Careful pelvic examination is necessary for diagnosis. Although USG itself is sufficient for diagnosis, MRI and IVP are other imaging techniques for diagnosis(6). Diagnosis is confirmed with drainage of hematocolpos or pyocolpos during septum resection operation. Resection of only transverse septum or both transverse and longitudinal septum can be preferred for treatment(5). Favorable reproductive results were reported after surgery. In conclusion; OHRIVA,uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis, is a rare syndrome. Careful evaluation of patients with complaint of cyclic abdominal pain, dysmenorrhea and abdominal mass shortafter menarce is necessary for diagnosis of syndrome.

References

1. Orazi C, Lucchetti MC, Schingo PM, Marchetti P, Ferro F. Herlyn-Werner-Wunderlich syndrome: uterus didelphys, blind hemivagina and ipsilateral renal agenesis. Sonographic and MR findings in 11 cases. Pediatric radiology. 2007;37(7):657-65. Epub 2007/05/16.

2. Candiani GB, Fedele L, Candiani M. Double uterus, blind hemivagina, and ipsilateral renal agenesis: 36 cases and long-term follow-up. Obstetrics and gynecology. 1997;90(1):26-32. Epub 1997/07/01.

3. Han BH, Park SB, Lee YJ, Lee KS, Lee YK. Uterus didelphys with blind hemivagina and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome) suspected on the presence of hydrocolpos on prenatal sonography. Journal of clinical ultrasound : JCU. 2013;41(6):380-2. Epub 2012/06/09.

4. Jindal G, Kachhawa S, Meena GL, Dhakar G. Uterus didelphys with unilateral obstructed hemivagina with hematometrocolpos and hematosalpinx with ipsilateral renal agenesis. Journal of human reproductive sciences. 2009;2(2):87-9. Epub 2009/11/03.

5. Gungor Ugurlucan F, Bastu E, Gulsen G, Kurek Eken M, Akhan SE. OHVIRA syndrome presenting with acute abdomen: a case report and review of the literature. Clinical imaging. 2013. Epub 2014/01/28.

6. Dhar H, Razek YA, Hamdi I. Uterus didelphys with obstructed right hemivagina, ipsilateral renal agenesis and right pyocolpos: a case report. Oman medical journal. 2011;26(6):447-50. Epub 2012/01/19.