Blind Hemivajina ile Birlikte Görülen Uterus Didelfis ve İpsilateral Renal Agenezisi Olan İki Olgu Sunumu

Uterus Didelphys with Unilateral Blind Hemivagina and Ipsilateral Renal Agenesis: Report of two cases

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Özet

Blind hemivajina ile birlikte görülen uterus didelfis ve ipsilateral renal agenezi normal menstruel siklusa sahip kadınlarda görülebilen nadir bir anomalidir. Bu olgu sunumunda çift uterus, unilateral vajinal obstrüksiyon ve ipsilateral renal agenezisi olan iki olgu raporlanmıştır. Her iki olguda pelvik ağrı, dismenore ve bununla ilişki olarak pelvik kitle tespit edilmiş virgin hastalardı. Bu sendromun farkına varılması ile vajinal septumun açılması reprodüktif kapasiteyi koruyarak semptomların tamamen düzelmesini sağlar.

Anahtar Kelimeler: Hematometra, Müllerian Anomali, Renal Agenezi
Türkçe Kısa Makale Başlığı: Blind Hemivajina

Abstract

Uterus didelphys with blind hemivagina and ipsilateral renal agenesis is a very rare anomaly and may be found in a female with normal menstrual periods.We report two cases with a double uterus, unilateral vaginal obstruction and ipsilateral renal agenesis. Both of the patients were virgin and their common clinical presentation was that of the onset of pelvic pain and dysmenorrhea, in association with the presence of a pelvic mass. In both of the patients we accurate the diagnosis by the laparotomic approach. A greater awareness of this syndrome should lead to accurate diagnosis and excision of the obstructing vaginal septum offers a complete relief of symptoms while preserving reproductive capacity.

Key words: *Hematometra, Mullerian Anomaly, RenalAgenesis*
İngilizce Kısa Makale Başlığı: *Blind Hemivagina*

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Introduction

Uterus didelphys with blind hemivagina and ipsilateral renal agenesis is a very rare anomaly and may be found in a female with normal menstrual periods (1-3).This is an uncommon anomaly of the development of the Mullerian ducts in which a defect in one of the Wolffian ducts leads to failed induction in kidney formation and in the fusion of the Mullerian ducts (4,5). Accurate diagnosis may be extremely difficult, because of the rarity of this lesion and unawareness of the condition(6).

Case reports

Thirteen and fifteen-years-old girls admitted to our clinic with pelvic pain. They were in the middle of their menstrual cycles and was not on regular menstrual periods. They were both virgin. The pain was episodic and colicky and aggravated around menstrual period especially for the last few months. On ultrasonographic examination uterine anomaly was suspected. In the 15 year old girl Computerized tomography (CT) yielded dermoid cyst in the non-cavitary cystic lesion of the uterus. She underwent laparotomy with suprapubic incision. Primarily bicornuate uterus with cystic mass at the middle was suspected at her operation and she referred our tertiarry center for further evaluation and operation. Her right kidney was not visualized at sonographic examination and intravenous pyelography revealed renal agenesia on the right side. Hematometra was diagnosed on her transrectal sonographic examination. Both adnexes were normal (Figure 1).

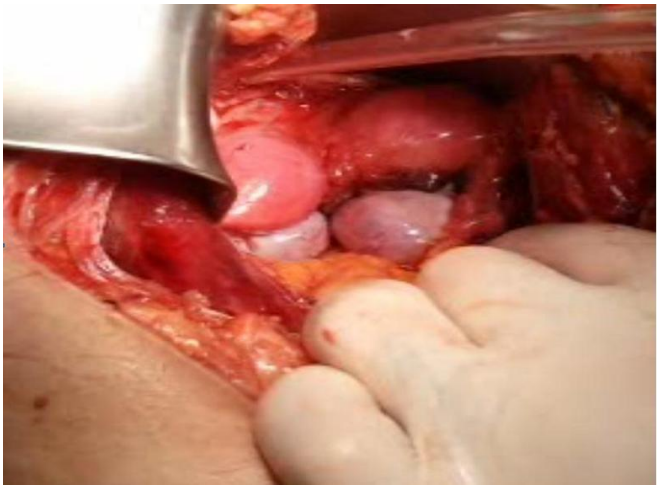

Figure 1: The appereance of uterus bicornis and bilateral overs

Hymenotomy was performed and vagina without cervix was located on the left side which was deviated and pushed by the hematometra mass on the right side. A longitudinal 3 cm incision was made on right vaginal lateral wall as much apicalas possible on the most prominent bulging area and thick viscous chocolate-like 1.5 liters of fluid drained through the incision (Figure 2).

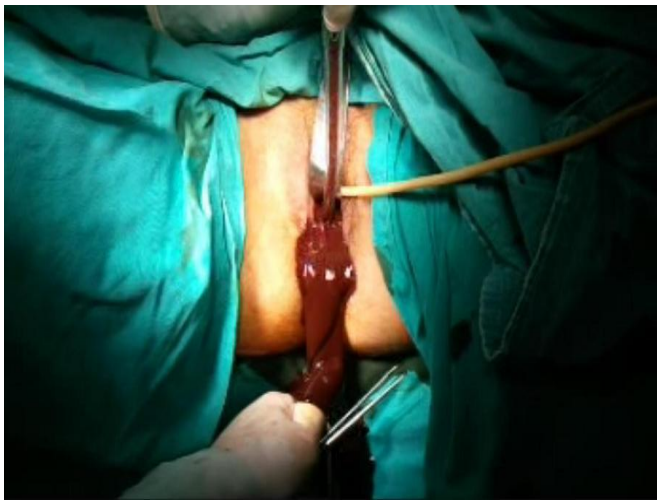

Figure 2 : Thick viscous chocolate-like 1.5 liters of fluid drained through the incision.

The cavity was washed with warm physiologic serum again and again until clear fluid was obtained. The incision was then marsupialized by interrupted no 3/0 polyglactin suture material. A foley catheter with 40 ml of fluid filled on its balloon was placed into the cavity to prevent closure of the incision. The patient at the age of 13 had the same clinical features except renal agenesis. The left hemivagina was filled by choclote like viscous fluid, hematometra and hematocolpos were identified. She has been operated by laparotomy and vaginal approach was applied as the same manner with the first patient. After the operations, both patients were regularly menstruating with no symptoms related with dysmenorrhea, endometriosis or pelvic pain. As the data related with the abnormal uterus can be collected only from a few case reports, this topic should be further discussed till etiopathogenesis is fully nlightened. Besides, rare occurrence makes its diagnosis a challenge for the gynecologists.

Discussion

Congenital abnormalities of the Müllerian duct system can result in various urogenital anomalies, including uterus didelphys with blind hemivagina and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome). Related to the precise relationship between renal agenesis and anomalies of the uterus during the developmental stage,the most accepted theory ,the fetal Wolffian ducts play an important role as a guide to the müllerian ducts to fuse in the midline. (4,5). If one of the Wolffian ducts is absent metanephric diverticulum, metanephric buds, metanephrogenic mesoderm, and finally one of the kidneys and collecting system can not develop. At the same time the mullerian duct, which is normally guided by the Wolfian duct, may fail to fuse together in the midline; hence a double uterus is formed and finally this displaced müllerian duct forms the imperforate hemivagina(2). Uterus didelphys is not a rare congenital malformation, but its association with unilateral imperforate vagina and ipsilateral renal agenesis is very rare. The accurate diagnosis may be difficult to make because the patient with a duplex Mullerian system and a unilateral intact vagina may have an abdominal mass and pain, although she may have normal menstruel periods.

An inexperienced physcian may misdiagnose such a case as either ovarian or abdominal pathology (7).

In one of our case was initially managed by another physician who failed to recognize the true nature of syndrome. Thus some of the patients underwent unnecessary or incomplete interventions. A rewiev of the literature revealed a few cases in which the preoperative diagnosis was accurate (7). However most of the cases were diagnosed during laparotomy or laparoscopy. Unfortunately with such a misdiagnosis, unnecessary surgical intervention may be done (8). When this anomaly is recognized laparotomy or laparoscopy should be avoided. When the accurate diagnosis is made intraoperatively, as in our cases, the intraabdominal intervention should be terminated. The correct treatment should consist of simple transvaginal septal excision (2,9,10). Our two patients were virgin. In both of the patiens, initially hymenotomi was performed and then the obstructed hemivagina was drained by making a vaginal incision. This was performed by the vaginal approach in both of the patients.

In conclusion, a greater awareness of the syndrome of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis should lead to its prompt diagnosis. This should allow for early and appropriate surgical intervention without compromising fertility.

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