Two Newborns Presenting with Intraabdominal Cystic Mass and Obstructive Uropathy: Hydrometrocolpos due to Vaginal Atresia

Karın İçi Kistik Kitle ve Obstrüktif Üropati ile Başvuran İki Yenidoğan: Vajinal Atreziye Bağlı Hidrometrokolpos

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Abstract

Vaginal atresia is a rare congenital anomaly and treatment approaches are varied, usually driven by symptoms. We report two newborns presenting with an intraabdominal cystic mass and obstructive uropathy due to vaginal atresia. Physical examination revealed a mass lesion extending to the umbilicus and lack of vaginal patency. They had polydactyly on hands and feet. Both patients underwent vaginoplasty with abdominoperineal approach. The patients are still in the 2nd postoperative year and their follow-up is uneventful. Vaginal atresia and hydrometrocolpos should be considered in patients with intraabdominal cystic mass and polydactyly findings.

Key Words: Vaginal Atresia, Obstructive Uropathy, Newborn

Introduction

Hydrometrocolpos is a rare malformation caused by accumulation of secretion in the uterine cavity and vagina due to congenital obstruction of the vagina (1). Vaginal obstruction may be result of various anomalies such as imperforate hymen, transverse vaginal septum, varying degrees of vaginal atresia or cloacal malformation (2). It is mostly detected during adolescence with symptoms of primary amenorrhea, cyclic abdominal pain and voiding dysfunction. Unusually, it may present itself with abdominal mass in the neonatal period and if large enough, may also show signs of urinary system partial obstruction for examples hydrourereters and hydronephrosis (2,3). Here, we report two newborns presenting with intraabdominal cystic mass and obstructive uropathy due to hydrometrocolpos secondary to vaginal atresia.

Case Presentation

Case 1

This infant was born to non-consanguineous healthy parents. The mother (G3, P2) was 21 years of age. The pregnancy was uneventful. On routine ultrasonography (USG) at 34 weeks a large cystic mass was found in the
fetal abdomen. The increasing size of the cyst on repeat examination led to early delivery by caesarean section at 36+0 weeks. A female child with weight 2870 grams was delivered with Apgar scores of 3 and 6 at 1 & 5 minutes. The newborn was intubated due to respiratory insufficiency and was immediately transferred to neonatal intensive care unit. The initial babygram showed a large abdominal mass displacing the intestine (Figure 1). Abdominal USG revealed bilateral grade 3 hydronephrosis and a complicated cystic lesion measuring 7x8 cm in the abdomen. Renal function tests were normal. Echocardiography was normal. Abdominal magnetic resonance imaging revealed complicated cystic appearance with preliminary diagnosis complicated mesenteric cyst which was approximately 7x8 cm in size, extending to the level of the superior umbilicus, filling the pelvic area completely (Figure 2). The patient was consulted to pediatric surgery department with the preliminary diagnosis of mesenteric cyst with these findings. Physical examination revealed a mass lesion extending to the umbilicus. Vaginal opening was not present. The appearance of the anus and urethral opening was normal. She had polydactyly on hands and feets. Surgery decision has been taken with the diagnosis of hydrometrocolpos due to vaginal atresia.

Case 2

A female baby with a birth weight of 3500 gr, was born by spontaneous vaginal delivery at 39 weeks of pregnancy from 34-year-old mother in her 4th pregnancy in the external center. The patient has been entubated due to respiratory insufficiency. Apgar score was unknown. The mother did not have regular antenatal care and no prenatal ultrasound examination was performed. The patient was referred to our hospital with evaluation of cystic mass on the abdominal USG which was done due to abdominal distension. Physical examination revealed abdominal distention and palpable mass extending to the superior of the umbilicus (Figure 3a). There was no vaginal patency (Figure 3b). The appearance of the anus and urethral opening was normal. She had polydactyly on hands and feets. Laboratory tests revealed that urea was 102 mg/dL (10.7-38.5) and creatinine was 2.01 mg/dL (0.57-0.87). USG revealed right grade 3, left grade 4 hydronephrosis and intra-abdominal cystic lesion measuring 10x11 cm. Echocardiography showed patent foramen ovale. Surgery decision has been taken with the diagnosis of hydrometrocolpos due to vaginal atresia.

Figure 1: The appearance of a pelvic mass that displacing the intestine on direct X-ray

Figure 2: The view of cystic uterus (asterisk) and hydronephotic kidney (arrow) on MRI

MRI: Magnetic resonance imaging
Both patients underwent vaginoplasty with abdominoperineal approach. Under general anesthesia with Pfannenstiel incision, an opening was made on anterior portion of the cystic uterus and mucoid fluid was aspirated. An urethral bougie was pushed from the uterus to vagina and vaginal bulging area was detected. The atretic vagina was reached with a cruciform incision made from this part, dissected and opened to the outside. Vaginal pull-through was completed (Figure 4a-b). For drainage, a 14 Fr silicone foley catheter was placed and the operation was terminated. Inserted catheter removed after six weeks and started daily dilatations later. Patients were seen monthly at the outpatient clinic. The case 2 did not come to the 5th month control. When she came to the 6th month control, it was difficult to apply bougie in her physical examination. It was learned that dilatation has not been performed for the last two months. The examination under general anesthesia was performed in 6 months postoperatively. Vaginal opening was found to be adequate in case 1. Case 2 was dilated under general anesthesia. Daily dilatations were continued after the procedure. Further, the hydroureronephrosis also regressed in time. The patients are still in the the 1st postoperative year and the follow-up is uneventful.

**Discussion**

The uterus and vagina are formed from the mullerian duct system which developed from an invagination of the coelomic epithelium. The fusion of distal portion of the mullerian duct forms the uterus and the proximal two third of the vagina. The lower third of the vagina is derived from the urogenital sinus which is the caudal ends of the Wolffian ducts. The development of the mullerian duct and genitourinary sinus affected by

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**Figure 3:** (a) Intraabdominal mass extending to the superior of the umbilicus. (b) View of closed vaginal entrance

**Figure 4:** (a) Operational view of urethral bougie that pushed from the uterus to vagina. (b) The appearence of new vaginal opening
various unidentified factors, and the different factors cause their development to stop at different stages, eventually caused to different types of vaginal dysplasia (4-6).

Gupta and Sharma (1) classified of vaginal dysplasia based on the level of obstruction and the severity of malformation: type I or low hymenal obstruction (Imperforate hymen); type II or mid-level obstruction (vaginal septum); type III or high obstruction (distal vaginal atresia); type IV due to urogenital sinus, and type V due to vaginal atresia with cloacal anomaly. In this article, we present two newborns with type III distal vaginal atresia.

Congenital vaginal atresia is a rare obstructive anomaly of the female genital tract. This generally occurs sporadically, with a reported incidence at term of 0.014-1%, and gives rise to hydrometrocolpos in less than 1/16,000 female births (7). Most cases are sporadic, though familial occurrence and an autosomal recessive mode of transmission (McKusick-Kaufman syndrome: hydrometrocolpos, postaxial polydactyly and cardiopathy) and (Bardet-Biedl syndrome: vaginal atresia, obesity, retinal pigmentation and mental retardation in adolescence) are reported (8,9). Our both patients had postaxial polydactyly but did not have cardiological anomaly or other stigmates suggesting these syndromes.

The presenting clinical features vary with age. The anomaly may stay undetected until adolescence, patient presents with primary amenorrhea or abdominal pain due to an obstructed uterovaginal tract. In the neonatal period, patients usually apply with abdominal mass and symptoms and complications related to the compression of mass (2). The differential diagnosis of lower abdominal masses in female neonates should include ovarian cyst, ureterocele, urachal cyst, anterior meningocele, mesenteric cyst, duplication of the bowel, giant urinoma, and megacystis (10). Additionally, associated genitourinary (persistent urogenital sinus) and anorectal anomalies (cloacal dysgenesis) may be included in the differential diagnosis (11).

Delay in diagnosis may cause several complications such as infection, sepsis, endometriosis and some other urogenital and cardiac complications related to compression (2). The perineum should be carefully examined to identify vaginal opening, imperforate hymen or a low transverse vaginal septum. For example, as in the first case presented, the patient was consulted with a mesenteric cyst pre-diagnosis. Unnecessary imaging studies and time loss may be avoided with a detailed physical examination. However, in some patients, it may not be clearly differentiated by external genital examination especially in urogenital sinus anomalies and cloacal malformations. Vaginoscopy may be helpful in determining the diagnosis and surgical strategy.

The management of patients with genital anomalies is a complex problem requiring individual surgical approaches, depending on the anatomical conditions. The level of the atresia is decisive in the surgical approach. In cases that very close to perineum, perineal repair becomes possible. But in cases with high atresia, firstly drainage is applied and abdominoperineal repair may be performed in a second session or drainage and abdominoperineal repair may be done in the same session. It is obvious that an obstructive vaginal anomaly should be treated surgically. However, the optimal timing for the surgical procedure is still in dispute. Shaked et al. (12) recommend performing emergency drainage in neonatal period and postponing definitive intervention postpone to adolescence. Both of the cases we presented had high level of vaginal atresia. In both cases, abdominoperineal repair was performed in a single session and no problem was encountered in long-term follow-up. The second important point to be considered in postoperative follow-up is regular dilatation program. Thus, partial stenosis was developed in case 2 due to regular dilatation was not performed.

Vaginal atresia is a rare congenital anomaly. Vaginal atresia and hydrometrocolpos should be considered in the differential diagnosis of female babies with a cystic mass in the abdomen during the intrauterine period. While the treatment options vary according to the level of atresia, in isolated vaginal atresia cases, abdominoperineal approach in single session is suitable option due to closer anatomical structures, flexibility of tissues in the neonatal period, in also high level cases. Postoperative dilatation program reduces complications. Considering the syndromes associated with vaginal atresia, the presence of additional anomalies such as polydactyly and congenital heart disease should be investigated.

**Ethics**

**Informed Consent:** Informed consent was obtained.

**Peer-review:** Externally peer-reviewed.

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**References**


