doi: 10.5505/abantmedj.2016. 25901

Abant Medical Journal

Orijinal Makale / *Case Report*

Volume Cilt 5 *Issue* Sayı 3 *Year* Yıl 2016

Posterosagital Anorektoplasti Sırasında Vajinal Anomalilere Dikkat

Attention of Vaginal Anomalies During the Posterosagittal Anorectoplasty

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Özet

Anorektal anomalili olgularda eşlik eden ürogenital anomali insidansı oldukça yüksektir. Rektovaginal fistüllü anal atrezili iki olgumuzda farklı şekilde karşımız çıkan vaginal anomali nedeniyle anorektal anomalili kızlarda olası vaginal patolojilere dikkat çekmek amacıyla olgularımız sunuldu.

Rektovestibuler fistüllü anal atrezi nedeniyle 8 aylıkken kliniğimize başvuran birinci olguda rektosigmoid kolostomi açıldı. Definitif ameliyatı sırasında iki vaginal orifis olduğu gözlendi. Posterosagittal anorektoplasti yapılırkenvaginal septum eksize edildi ve vaginoplasti gerçekleştirildi. Yenidoğan döneminde rektovestibuler fistüllü anal atrezi nedeniyle sol sigmoid kolostomi yapılan ikinci olguda definitif ameliyat sırasında vaginal orifis izlenmedi, ameliyata son verildi. Hastanın kromozom analizinde 46XX olduğu, Pelvik MRI incelemede atrofik uterus olabilecek şüpheli imaj olması nedeniyle vaginal atrezi ön tanısı konuldu. Hastaya 2 yaşında posterosagittal yaklaşımla mevcut rektovestibüler fistülden neovagina oluşturularak anorektovaginoplasti yapıldı.

Sonuç olarak; rektovestibüler fistüllü anal atrezili kızlarda eşlik edebilecek vaginal anomaliler açısından hastanın ilk muayenesinde perinenin dikkatlice gözden geçirilmesi; definitif ameliyatlar sırasında vaginal anomali sürprizleriyle karşılaşılmasını azaltacaktır.

Anahtar Kelimeler: Vaginal anomali, vaginal atrezi, anorektal malformasyonlar, kloaka anomalisi, rektovestibular fistül.

Abstract

The incidence of urogenital anomaly associated with anorectal anomalies is very high. Two different types of vaginal anomaly associated with anorectal anomalies were presented in here.

An 8 months old girl admitted to our department due to anal atresia with rectovestibular fistula. A rectosigmoid colostomy was created due to very narrow fistula. During the definitive operation, two vaginal orifices were seen; vaginal septum excision and vaginoplasty via posterosagital anorectoplasty were done. Another girl admitted to our department anal atresia with rectovestibuler fistula who had one year old. Calibration and dilatation of the fistula had been done until Hegar 15. Patient had been prepared for definitive operation without colostomy; but vaginal orifice was not found in perineal examination and a diverting sigmoid colostomy was created. Chromosomal analysis was found as 46 XX. Prediagnosis was vaginal atresia, because pelvic MR showed a suspicious imagination like an atrophic uterus. Vaginal orifice was absent at cystoscopy. Bifid uterus, right and left tubas, and right and left ovaries were seen at laparoscopy. A neovagina from rectovestibular fistula during the posterosagittal anorectoplasty was created in two years old, and three months later colostomy closed.

In conclusion; girls with anorectal anomaly should be examined carefully at the first admission to determine the associated vaginal anomalies and to avoid surprises during the surgery.

Keywords: Vaginal anomaly, vaginal atresia, anorectal malformations, persistent cloaca, rectovestibular fistula.

INTRODUCTION

The incidence of vaginal anomalies in infants is very rare and the etiology of vaginal anomaly is unknown. They can differ from vaginal agenesis to double vagina. Pure vaginal anomaly is a very rare condition, however some syndromes and/or anorectal anomalies can accompany the vaginal anomalies (1,2,3,4).

Vaginal atresia is estimated to occur in 1 in 4000–5000 live female births. It can stay often unnoticed until adolescence period, when pain and a lack of menstrual flow indicates these condition. When a vaginal atresia is diagnosed, there are numerous treatment methods based on the exact details of the pathology. In some cases, a new vagina can be reconstructed using an intestinal graft (1,2,3,4). Herein we present two different rare types of vaginal anomaly associated with anorectal malformations.

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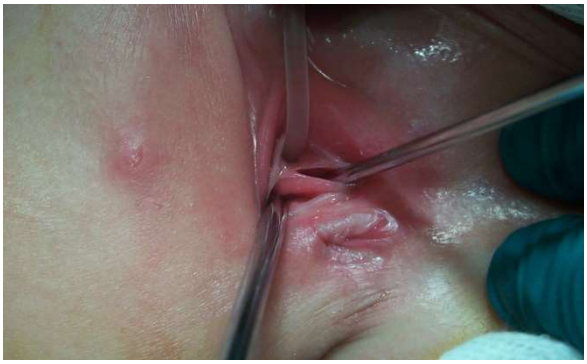
Geliş tarihi / *Received*: 21.08.2015 **Kabul tarihi / Accepted:** 16.02.2016 **Çıkar Çatışması / Conflict of Interest:** Yok / None

CASE REPORT:

Case 1: An 8 months old girl admitted to our department due to anal atresia with rectovestibuler fistula. She also had 4. and 5. sacral vertebral deformities. A diverting rectosigmoid colostomy was created due to very narrow fistula. During the definitive operation, a foley catheter was inserted to the urethra, and two orifices of vagina were identified (Figure 1); and two columns of uterus were seen endoscopically. Vaginal septum excision and vaginoplasty via posterosagital anorectoplasty were performed. Colostomy were closed after the dilatation programme of neoanus.

Figure 1: Two vaginal introitus of double vagina is seen in case one. Arrow shows rectovestibuler fistula.



Case 2: One year old girl admitted to our department with anal atresia with rectovestibuler fistula. Calibration and dilatation of the fistula had been done to number 15 Hegar and the patient prepared for definitive operation without colostomy. However during the operation vaginal orifice was not seen in perineal examination and a diverting sigmoid colostomy was performed (Figure 2). Choromosomal analysis was found as 46 XX. Prediagnosis was vaginal atresia, because pelvic MRI showed an suspicious imagination like an atrophic uterus. She also had severe lumbosacral deformity. Vaginal orifice was absent at cystoscopy. Bifid uterus, right and left tubas, and right and left ovaries were seen at laparoscopy. A neovagina from rectovestibular fistula during the posterosagittal anorectoplasty was created at two years of age, and three months later colostomy was closed.

DISCUSSION

The most common clinical presentation of vaginal anomalies are associated with anorectal malformations. Clinical presentation of this association is the same as that for any anorectal malformation and vaginal anomaly in the neonatal period. Thus, it is difficult to diagnose this association in the neonatal period. The vaginal anomaly is easy to confuse on a physical exam as it mimics the anorectal malformation. Sometime vaginal anomalies can be diagnosed intraoperatively especially during posterior sagital anorectoplasty (5)

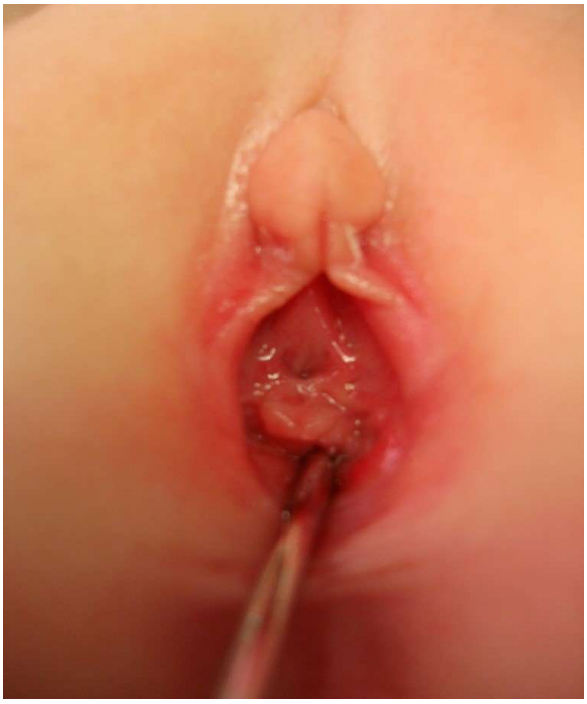
It is well known that abnormalities of vagina are rare but common in anorectal malformations; and are found in patients following careful examination at admission (2,5). Accordingly, spinal ultrasonography or magnetic resonance imaging should be performed in patients with anorectal malformations to rule out vaginal pathology such as atresia, and agenesis, or duplication. Such conditions can mistakenly be attributed to the reconstructive procedure of the anorectum (2,5-8).

The radiological differential diagnosis for vaginal anomalies is possible, but first it must be thought. The diagnosis of vaginal anomalies is usually made following initial administration or during the operation for anorectal anomalies (5). The main features that help distinguish vaginal anomalies are with the advent of modern minimally invasive endourology techniques. A carefully examination of the perineum is essential in the diagnosis of vaginal anomalies.

Besides a physical examination, the physician will need imaging techniques such as gynecologic ultrasonography, pelvic MRI, or hysterosalpingography to determine the character of the malformation. A hysterosalpingogram is not considered as useful

due to the inability of the technique to evaluate the exterior contour of the uterus and distinguish between a bicornuate and septate uterus in newborn and children. In addition, laparoscopy and/or hysteroscopy may be indicated (5).

Figure 2: Vaginal introitus is seen as closed under the urethral orifice (arrow shows urethral orifice) in second case. Hegar dilatator is entered into the rectovestibular fistula.



The first procedure involves a careful examination followed by using a flexible ureteroscope. Upon confirmation of the diagnosis the patient is then listed for a second stage operation of the vaginal anomalies according to the etiology. It is essential to delineate the anatomy of the vaginal anomaly using the available preoperative images. With the widespread availability of multiphase CT scanners, allowing imaging during phases of contrast excretion and capacity of coronal reconstruction, CT images may be satisfactory (5). Under general anesthesia a flexible ureteroscope or rigid sistoscope is performed to delineate the anatomy of the urogenitalia (2).

Indeed, double vagina has been identified as the first case easily. Double vagina is possible to determine by careful examination of the perineum; but, detection of vaginal atresia is usually not easy. Although, based on these results, a simple vaginal atresia was unlikely, the presence of some syndromes like Herlyn-Werner-Wunderlich syndrome was excluded by chromosomal analyses, cystoscopy, laparoscopy, and MRI in second case. Following detailed examination under the general anesthesia, cystoscopy and laparoscopy performed prior the definitive operation of anorectal malformations in second case.

In conclusion, pure vaginal anomalies are rare lesions but the possibility of vaginal abnormality should be considered in patients with anorectal malformations. Careful inspection of the perineum after general examination of the patient had not reveal any significant misdiagnosis or late diagnosis. Radiological investigations using contrast materials followed by cystoscopy/laparoscopy are effective diagnostic strategies which prevents last minute surprises at operations in girls with anorectal malformation.

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