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A very rare type of cloacal malformation associated with duplication of appendix vermiformis

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Özet

Apandiks vermiformis duplikasyonu ile birlikte çok nadir bir kloakal malformasyon olgusu

Persisten ürogenital sinüsle birlikte uterus bicornusu, mesaneye açılan kısa kolonu ve çift apandiksi olan nadir bir kloakal malformasyon vakasını sunuyoruz. İlaveten, bu vakada MR çalışması ile spinal deformite ve tethered kord da saptanmıştır. Kloakanın sık gözlenen formlarına göre bu tip anomaliye yaklaşım seçenekleri çok daha sınırlı kalmıştır.

Anahtar kelimeler: *Kloakal malformasyon, çift apandiks vermiformis*

Summary

We present an unusual case of cloacal malformation where the persistent urogenital sinus is associated with uterus bicornus, a short colon opening into the bladder and double appendix vermiformis. Additionally, MRI investigation revealed spinal deformity and tethered cord. Management options for this type of anomaly are more restricted than the common form of cloaca.

Key words: *Cloacal malformation, double appendix vermiformis*

Introduction

Cloacal malformations and duplication of appendix vermiformis are two extremely rare anomalies with incidences of 1 in 50000 and 2 in 50000, respectively [2,3]. Individual cases of cloacal malformations are peculiar in certain aspects and must be defined appropriately.

On the other hand, although the duplication of appendix is an incidental finding in abdominal surgery, but when detected in childhood almost all patients are noticed to have associated intestinal or genitourinary malformations [1].

We present a case of cloacal malformation with spinal abnormalities associated with type- A duplication of appendix which is a rare association in this spectrum of anomalies.

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Case Report

A baby of female phenotype was referred to our clinic for abdominal distention and vomiting. She was found to have a single perineal opening with imperforate anus and her abdominal distention appeared to be related to a mass. Ultrasound examination demonstrated a suprapubic cystic lesion, 8x6x6 cm in size and bilateral mild hydronephrosis. It was possible to decompress this cyst by passage of a catheter through the perineal opening. The discharge of urine and meconium was indicative of cloacal malformation.

The patient was subsequently operated and found to have a short malformed colon communicating with the bladder. The uterus was bicornuate and the cecum carried two vermiform appendices (Figure 1). The colonic fistula to the bladder was divided and exteriorized as an end colostomy. The bladder itself was repaired, because a vesicostomy was not thought to be necessary.

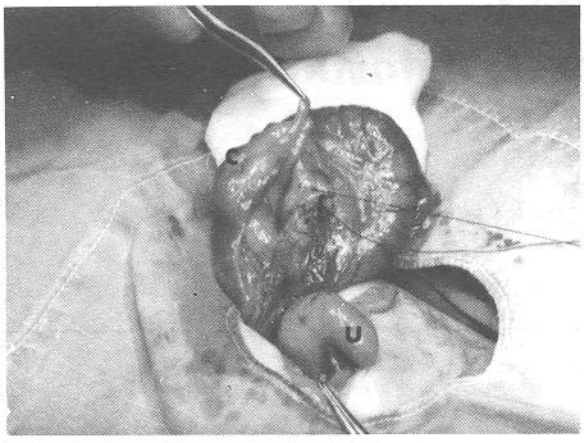


Figure 1. Appendiceal duplication with single cecum (c) and one of the uteri (u) are seen. "f" indicates the fistula.

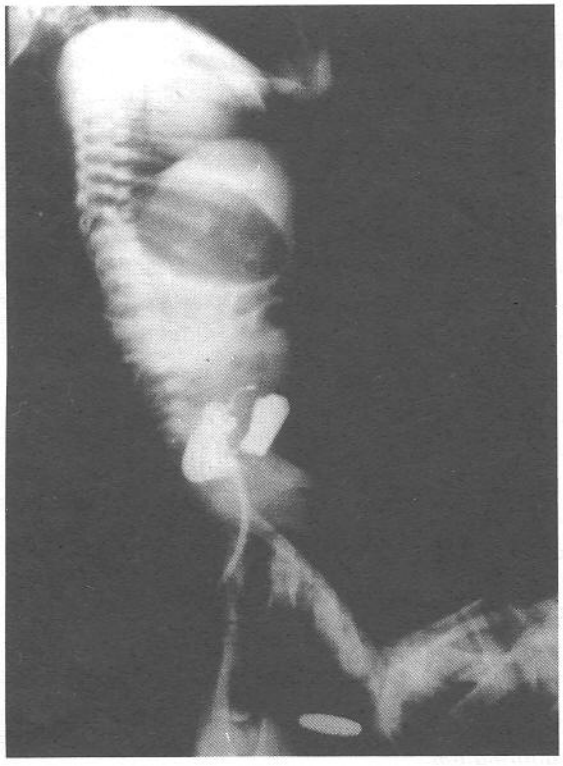
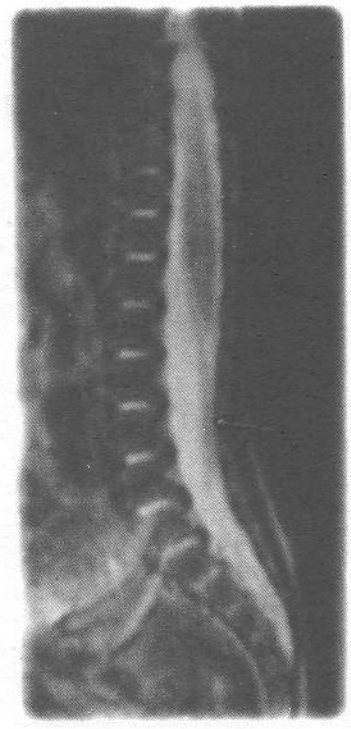


Figure 2. Sinogram showing a large uterovaginal cavity posteriorly and bladder anteriorly.

Sinogram and magnetic resonance imaging (MRI) studies were performed postoperatively. With sinogram, a large uterovaginal cavity and a bladder with unilateral vesico-ureteral reflux were shown (Figure 2). With MRI study, enlargement of spinal canal and tethered cord were found and pelvic anatomy was delineated (Figure 3 a,b).



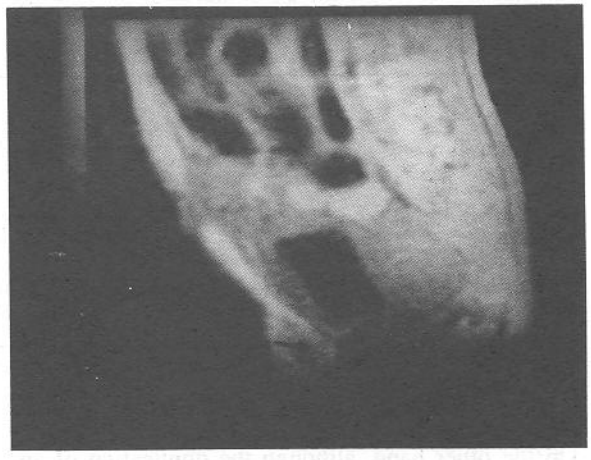


Figure 3 a) MRI study obtained postoperatively revealing tethering cord (arrow), b) pelvic structures.

It was decided to give priority to the reconstruction of the lower urinary tract. The patient is currently symptom-free and can void spontaneously.

Discussion

A cloacal malformation denotes the confluence of the urinary, genital and gastrointestinal tracts in the female patient [14]. Cloacal anomalies are very complex anatomic structures but the basic form is

consistent. Gastrointestinal tract opening to the bladder is a very rare type of cloacal malformation. Pena, in his series, classified the type of cloacal malformation with rectal opening in bladder as an unusual variant [7].

Our patient had a very high anorectal agenesis at approximately 15 cm distal to cecum in addition to this very rare type of opening. Genital anomalies such as duplications, urinary and vertebral anomalies are common associated malformations in these children.

In 1962, three types of duplication of appendix have been described: type A, partial duplication of appendix to various degrees on a single cecum; type B, a single cecum with two completely separate appendices (B1: the bird-like type and B2: the tenia coli-type); type C, double cecum with each limb bearing an appendix and most cases are the bird-like type [8,9].

Type B1 and type C of duplications of appendix are usually together with other intestinal, genito-urinary and spinal anomalies, while the type A and B2 duplications of appendix have no other associated anomaly [6].

Our case was a typical type A duplication of appendix that is a single based appendix with partial duplication. Type A anomaly is believed to occur by fusion of the transient appendix with the precursor of the normal appendix during embryological development [6].

Intestinal and genitourinary septum anomalies coincide with appendiceal duplications because of the insufficiency in cloacal differentiation [8]. But in the literature we could not find any type A appendiceal duplication associated with other congenital abnormalities.

Finally, since the coincidence of the anomalies of the lower spinal cord is very high with cloacal malformations, the evaluation of this anatomic region with MRI is essential especially to rule out tethered cord which could have functional consequences.

MRI would seem to be a usefull screening method in the search of lumbosacral spinal cord and genitourinay lesions although plain radiography of the spine, ultrasound and cystography will continue to play an important role in the planning of long-term management of these patients [5].

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