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Functional medicine

A rare presentation: Penile agenesis, vesicoureteral reflux, and rectovesical fistula in a newborn☆

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ABSTRACT

The penis is an important organ in fertility, urinary and psychosexual structure of males. Complete penile agenesis is a rare congenital genitourinary anomaly and is usually associated with other malformations such as gastrointestinal, cardiac, and musculoskeletal lesions. Although many uncommon types have been reported, penile agenesis associated with vesicoureteral reflux has not been reported in an infant with vesicorectal fistula. This is the first report of an infant with the combination of penile agenesis, vesicoureteral reflux, and vesicorectal fistula. A three-year-old infant with penile agenesis proven to have bilateral vesicoureteral reflux and vesicorectal fistula is reported.

Introduction

Penile agenesis (PA) is a rare penile congenital abnormality, and its incidence is one in 10–30 million live births. 80 PA cases have been reported in the literature until now.[1] The occurrence of genitourinary anomalies has been reported in association with numerous pathologic alterations classified as vertebral/skeletal, gastrointestinal, and miscellaneous. The combination of symptoms is non-random. These malformations are presumed to result either due to disturbed migration of the genital tubercle or its failure to develop and to originate somewhere the 12th weeks of gestation during early embryogenesis.[2] Although a combination of symptoms associated with genitourinary anomalies and anorectal malformation (ARM) has been reported,[3–5] there is no report of the penile agenesis (PA) associated with bilateral vesicoureteral reflux (VUR) and vesicorectal fistula (VRF) in an infant. We present this case of congenital aphallia for which early diagnosis and treatment is very important to help the infant and its family.

Case report

A 1-day-old male newborn was admitted to the Necmettin Erbakan University Meram Medical Faculty Department of Pediatric Surgery with the diagnosis of PA (Fig. 1). The patient had a 46 XY karyotype, bilateral descended testes in well-formed scrotums. Cystostomy operation was performed after a physical examination and radiographic investigations on the same day. PA has not been reported in an infant with vesicorectal fistula (VRF) and unilateral grade 2 vesicoureteral reflux (VUR). Bilateral vesicoureteral reflux was identified after ligation of vesicorectal fistula in the patient (Fig. 2). No other associated anomalies were identified. During the surgery, diagnostic laparatomy and colostomy were performed in newborn period. Ureteral orifices could not be seen, but 2-3 cm-long anatomic body such as urtethra, which could be identified with 3 Fr ureteral cathater, was seen in cystoscopy at the age of 4 months. Vesicorectal fistula ligation was carried out when he was 8-months-old. 3 weeks later, his colostomy was closed. Endoscopic treatment could not be performed because ureteral orifices could not be identified in cystoscopic examination at the age of 1 year. But endoscopic subtrigonal injections were performed at the age of 4 year. Bilateral ureteroneocystostomy in sufficient bladder capacity and penile reconstruction, which permits clean intermittent catheterisation, in adolescence period were scheduled. 8 Fr cystostomy cathater was settled into the bladder for diuresis. Postoperative evaluation documented no stone, infection, and malignancy in the bladder. Fecal continance of the patient, who had fecal incontinence, was carried out with intestinal management programme. The patient followed with 8 Fr cystostomy cathater and cefixim prophylaxis is 5-years-old, he has been following up at 1 month intervals and his kidney functions are good (urea: 35.7 mg/dl and creatinine: 0.56 mg/dl).

Discussion

The incidence of penile agenesis (PA) has been estimated to be 1 in

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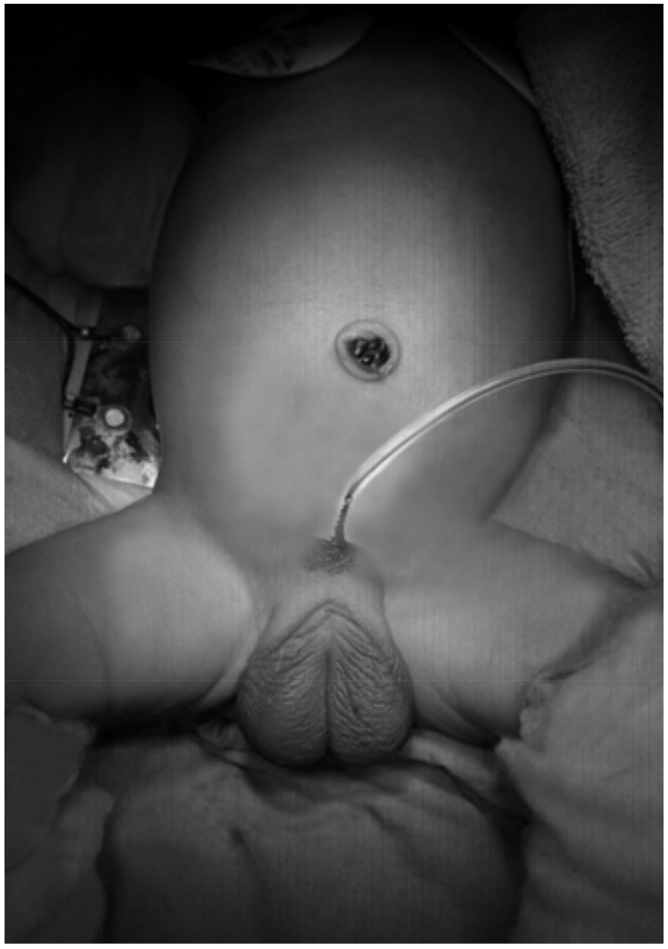

Fig. 1. The appearance of the congenital aphallia in a newborn. Note the absence of penis and urethra despite bilaterally descended testes.

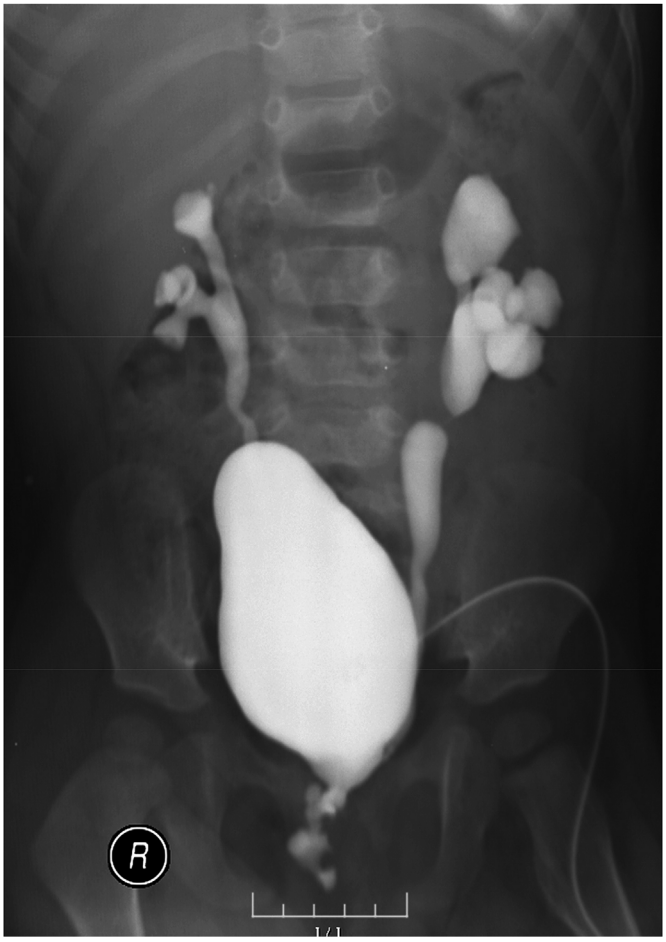

Fig. 2. The appearance of the bilateral vesicoureteral reflux (after ligation of rectovesical fistula).

10–30 million live births, whereas PA has never been reported in the literature.[1] Variants of PA may include cryptorchidism, renal anomalies, vesicoureteral reflux, anal anomalies, and cardiac anomalies. Female gender reassignment with orchiectomy, along with urinary tract and genital reconstruction, has traditionally been recommended for this anomaly. However, an entire intersex assessment team, including pediatric urology, endocrinology, and psychiatry was formed before full informed consent was obtained from the family.[1,5]

There is evidence that the pathogenesis of PA and genitourinary anomalies are similar. It is obvious that these malformations are presumed to result them. These malformations are presumed to result either due to disturbed migration of the genital tubercle or its failure to develop and to originate somewhere between the 12th weeks of gestation during embryogenesis.[2] Although this theory may explain the pathogenesis of PA, our case supports the option of a malformation occurring during early pregnancy.[2]

The associated anomalies in our case with PA, bilateral VUR, and VRF reinforce the theory of a malformation process such as a disturbed migration of the genital tubercle or its failure to development taking place before the 12th week of embryonic life.[2] Our case is a new example of an associated malformation in the newborn with PA and raises the discussion of possible etiologic factors. To the best of our knowledge and following a review of the PubMed database, we report a never before described associated anomaly with PA making this report a unique contribution to the world literature. This may show that a solitary explanation for the pathogenesis is difficult to define. The one possibility of a disturbed migration of the genital tubercle or its failure to develop will likely answer the questions concerning the etiology of both PA, VUR, and VRF.[1]

8 Fr cystostomy cathater settled into the bladder for diuresis may be an alternative to the conventional procedure. We performed the perineal fistula ligation after cystostomy cathater settlement in this case to prevent urinary tract infection. We think that the penile reconstruction will be a good alternative for this patient with bilateral VUR, and VRF even though it is rarely seen. Thus, an artificial penile conduit may be necessary later as a urethral tract reconstruction in newborns with bilateral VUR and VRF.

Conflicts of interest

There is no conflicts of interest.

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