

Case Report

Posterior urethral valve and ureterocele in a neonate: A rare association

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ABSTRACT

Background: The posterior urethral valve and ureterocele are usually present as two separate entities in neonates. Their coexistence appears to be extremely rare owing to different embryonic developmental defects.

Case Presentation: We herein present this rare association in 3 weeks old male with antenatal right hydroureteronephrosis. The patient underwent cystoscopy with fulguration of the posterior urethral valve along with endoscopic drainage of ureterocele and was doing well on follow-up for the last 5 years.

Conclusion: The authors discussed the management of this rare association to highlight the need for a systematic management approach.

Keywords: Posterior urethral valve, Ureterocele, Duplex kidney

INTRODUCTION

The posterior urethral valve is one of the most common causes of lower urinary tract obstruction in neonates and usually presents as an isolated anomaly. [1-4] The development of the ureterocele in presence of a posterior urethral valve appears to be extremely uncommon owing to different embryological development and the presence of a high-pressure bladder. [1,2,4] The simultaneous occurrence of ureterocele and posterior urethral valves has no embryological linkage and should be construed as unrelated events. [1,2] The present case appears to be the third case of this rare association to be reported in English literature to date.

CASE REPORT

A 2-week-old male presented as a case of antenatal right hydronephrosis diagnosed at 27 weeks of gestation with no history of oligohydramnios during the gestational period. Post-natal ultrasound scan on day 3 of life confirmed diagnosis of right hydroureteronephrosis with normal left kidney and urinary bladder. Clinical examination showed normal external genitalia with non-palpable both kidneys in absence of any dysmorphic features. Laboratory investigations showed normal renal function tests with a serum creatinine of 0.2 mg/dl. Mic-

turating cystourethrogram showed right duplex kidney with grade 4 reflux in lower moiety along with right ureterocele. The urinary bladder appeared regular and smooth in outline with a non-dilated posterior urethra although the child passed urine in weak stream during voiding. So, the child underwent cystourethroscopy at age of 3 weeks which confirmed the presence of a right duplex kidney with a normally placed lower moiety ureteric orifice, the presence of orthotopic ureterocele in relation to an upper moiety, the presence of slightly trabeculated bladder and posterior urethral valve. The patient underwent an endoscopic puncture of the ureterocele with cold knife fulguration of the posterior urethral valve. On follow up patient remained free from urinary tract infection and was passing urine in good stream. MAG 3 renogram on follow-up showed a split renal function of 48% in right and 52% in the left kidney with the non-obstructive curve. On follow-up for 5 years patient remained continent for urine, free from urinary tract infection with improvement in radiological status.

DISCUSSION

Among congenital anomalies of the kidney and urinary tract (CAKUT) posterior urethral valve remains the most common cause of lower urinary tract obstruction in

newborn males with a reported incidence of 1 in 4000 to 8000 pregnancies. [1-4] Usually, the posterior urethral valve presents as an isolated developmental renal anomaly and rarely presents in association with renal anomalies like the Unicaiceal kidney, partial urethral duplication, and scaphoid megalourethra. [5-7] A review of the literature suggests that cryptorchidism remains the most common and ureterocele remains the rarest associated anomaly in a patient with a posterior urethral valve. [1,7]

The simultaneous occurrence of orthotopic ureterocele and posterior urethral valves has neither embryological nor physiological linkage. [1] The synchronous presence of ureterocele in presence of a high-pressure bladder due to the posterior urethral valve is difficult to explain physiologically. [1] Embryologically posterior urethral valve develops as a result of abnormal insertion and migration of the orifice of the mesonephric duct whereas formation and migration of the ureteric bud and its incorporation in the bladder resulting in obstruction to urine flow remain the embryological basis for ureterocele development. [3,4] Since both entities have different embryogenesis so the simultaneous occurrence of posterior urethral valve and ureterocele appears to be unrelated events and incidental in nature. [1,2]

Although the clinical presentation of both entities remains voiding difficulty remains with the posterior urethral valve presenting in infancy and ureterocele commonly presenting in older children; still, their

association lacks typical diagnostic presentation. [1] As experienced in present and reported cases patient usually presents with voiding difficulty and either of the associated anomalies are diagnosed as an incidental finding on micturating cystourethrogram. [1,2,7,8]

As experienced in present and reported cases endoscopic intervention in form of simultaneous puncture of ureterocele and fulguration of the posterior urethral valve remains the treatment of choice [1,2]. Both conditions require a systematic approach with a possible requirement of multiple procedures to avoid complications. As experienced in one of the reported cases, one of the pathologies can be missed owing to the rarity of association [2]. So, an insight among endourologist would facilitate prompt diagnosis and proper management of this rare association by vigilant endoscopic assessment and subsequent treatment to optimize renal statu.

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