A Rare Case of Orthotopic Ureterocele with Posterior Urethral Valve

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Abstract. Posterior urethral valves (PUV) and ureterocele are two entities with different embryological basis. The occurrence of these two conditions in one patient is very rare. We report a case of left side ureterocele and PUV during follow-up of a case of antenatally diagnosed left hydroureteronephrosis. Cystoscopy with deroofing of large ureterocele and incision of valves was done at 5, 7 and 12 ’O clock positions. The patient was asymptomatic and normotensive during last follow-up at 7 years of age. The development of ureterocele despite high bladder pressures associated with PUV is interesting. A systematic approach is needed to deal with such cases.

Keywords: Hydroureteronephrosis, Deroofing, Posterior urethral valve, Ureterocele, Valve incision

Introduction

Routine use of antenatal ultrasonography (USG) has lead to early diagnosis of various urological anomalies, including the detection of multiple anomalies. While most of these multiple anomalies can be theorized to have some embryological basis, we can rarely have associations which are purely incidental. One of these is a rare association of posterior urethral valve (PUV) and ureterocele.

Case report

Our case was a case of antenatally diagnosed left hydroureteronephrosis and was asymptomatic postnatally. Postnatal evaluation by USG and micturating cystourethrogram (MCU) revealed left hydroureteronephrosis with ureterocele and no vesicoureteric reflux (VUR). Cystoscopy and deroofing was done at 2 months of age in another hospital. Post-operative Diethylene Triamine Pentacaetic Acid (DTPA) scan revealed practically non-functioning left kidney with 4 % function (6 months age). Repeat USG (Fig. 1) and MCU (Fig. 2) at 5 years of age revealed left ureterocele, left grade 5 VUR and dilated posterior urethra with non-visualisation of left kidney on USG. Repeat cystoscopy with deroofing of large left ureterocele and incision of posterior urethral valves was done at 5, 7 and 12 ’O clock positions. Postoperative MCU (Fig. 3) after 1 year revealed near normal posterior urethra and left grade 3 VUR. The patient was asymptomatic and normotensive during last follow-up at 7 years of age.

Discussion

Posterior urethral valves (PUV) are mostly encountered in isolation and they rarely form a part of a conglomerate of congenital lesions. To the best of our knowledge in English literature, our case is the second case after Chawla et al[1] showing the occurrence of PUV and ureterocele in one patient. Posterior urethral valves (PUV) and ureterocele are two
entities with different embryological basis. PUV can have varied presentations ranging from antenatal diagnosis, oligohydramnios, hydrops to voiding symptoms since birth to mild symptoms in older children. Ureteroceles can also present with antenatal diagnosis, voiding symptoms in postnatal period to incidental diagnosis in adults. PUV is a disease of males while ureteroceles are more common in females (80%). The association of other urological anomalies like unicaliceal kidney, partial urethral duplication and scaphoid megalourethra in a case of PUV is relatively rare.\textsuperscript{[2-5]} Cryptorchidism forms the most common associated congenital anomaly with posterior urethral valves.\textsuperscript{[6]} Clinical expression of orthotopic ureterocele in infancy is extremely rare and the majority of symptomatic ureteroceles commonly involve upper pole ureters of duplex systems (80%).\textsuperscript{[7]} Both conditions require systematic approach with possible requirement of multiple procedures to avoid complications.

The development of ureterocele despite high bladder pressures associated with PUV is interesting. Voiding disability is a common symptom of both ureterocele and posterior urethral valves and hence a MCU to highlight unsuspected PUV as exemplified by this case report is indicated. While long term prognosis of PUV is guarded, the outcome of properly managed ureteroceles is usually excellent. A systematic approach is needed to deal with such cases.

References


