Journal of Pediatric Urology (2011) 7, 491—494

Journal of Pediatric urology

CASE REPORT

Segmental multicystic dysplastic kidney: A rare situation

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Received 13 July 2010; accepted 3 December 2010
Available online 5 February 2011

KEYWORDS
Multicystic dysplastic kidney

Abstract Segmental multicystic dysplastic kidney is a rare subtype, found in only about 4% of children diagnosed with MCDK. To the best of our knowledge, we describe the 36th reported case of segmental multicystic kidney disease.

Multicystic dysplastic kidney (MCDK) is a form of renal dysplasia in which there are multiple non-communicating cysts of various sizes separated by dysplastic parenchyma, resulting from abnormal and incomplete kidney development, and renal function is minimal or absent. Segmental MCDK is a rare subtype of this disorder, found in only about 4% of children diagnosed with MCDK. To the best of our knowledge, we describe the 36th case of segmental multicystic kidney disease [1—3].

Case report

An 8-year-old female presented after one episode of pyelonephritis and sepsis with a painless, 5-cm palpable mass in the left flank. Ultrasonography revealed the left kidney to be enlarged with multiple cysts affecting only the lower pole, no parenchyma detected, and clear definition of the cyst walls and no communication among them (Fig. 1A and B). The right kidney was normal. Computerized tomography (CT) showed a multiseptated cystic mass on the lower half of the left kidney which was evident in the non-contrast phase of the exam (Fig. 2A and B). The CT scan in the contrast phase indicated complete absence of function on the lower pole of the left side which was confirmed by DMSA scan (Fig. 3A and B). The DMSA renal scan revealed relative function of 44% in the left and 56% in the right, and the presence of scars could be identified (Fig. 4). Voiding cystourethrography (VCUG) showed bilateral vesicoureteral Grade IV—V reflux (VUR) and left duplex system with a blind-ended unit up to the mid-ureter correspondent to the dysplastic lower pole (Fig. 5A). A contrast CT scan image is shown which confirms the VCUG findings (Fig. 5B).

We performed a common sheath ureteral reimplantation (Cohen) and hemi-nephrectomy of the lower dysplastic pole. The child presented good clinical recovery.

Discussion

Segmental multicystic dysplastic kidney (SMDK) occurs either from insufficient interaction of the ureteral bud and

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1477-5131/$36
doi:10.1016/j.jpurol.2010.12.005

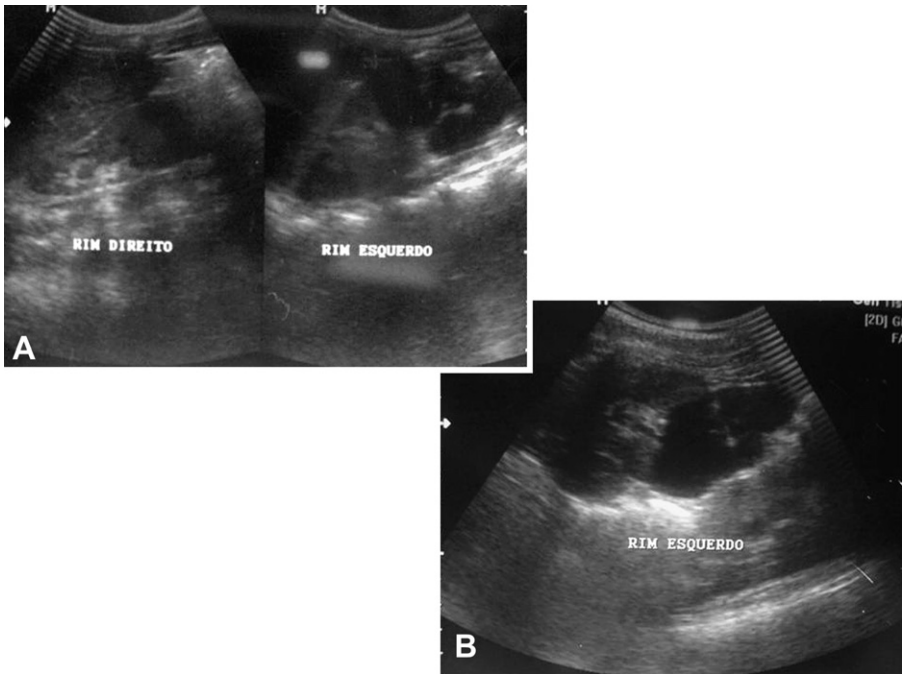

Figure 1 Ultrasonography reveals multiple cysts affecting only the lower pole of the left kidney.

metanephrons or from prenatal obstruction leading to cystic dysplasia. Cytomegalovirus infection has also been implicated in the development of renal dysplasia [2]. Genetics can also play a role, mainly when there is an association with VUR [4].

Thirty-five SMDK cases have been previously described. Such studies showed that the left kidney and females are mostly affected. These data reinforce the significance of genetics in SMDK development. As for associated anomalies, the presence of VUR in the contralateral kidney is found in 20—33% of cases. Regarding the ipsilateral kidney, the incidence of associated changes is 77%, lower pole VUR being the most common. These figures reinforce the need to evaluate the entire urinary tract before deciding on treatment [1].

Most of the reported cases were associated with duplex collecting systems. Only 6 of the 35 had a single collecting system [3]. This suggests a similarity between SMDK and

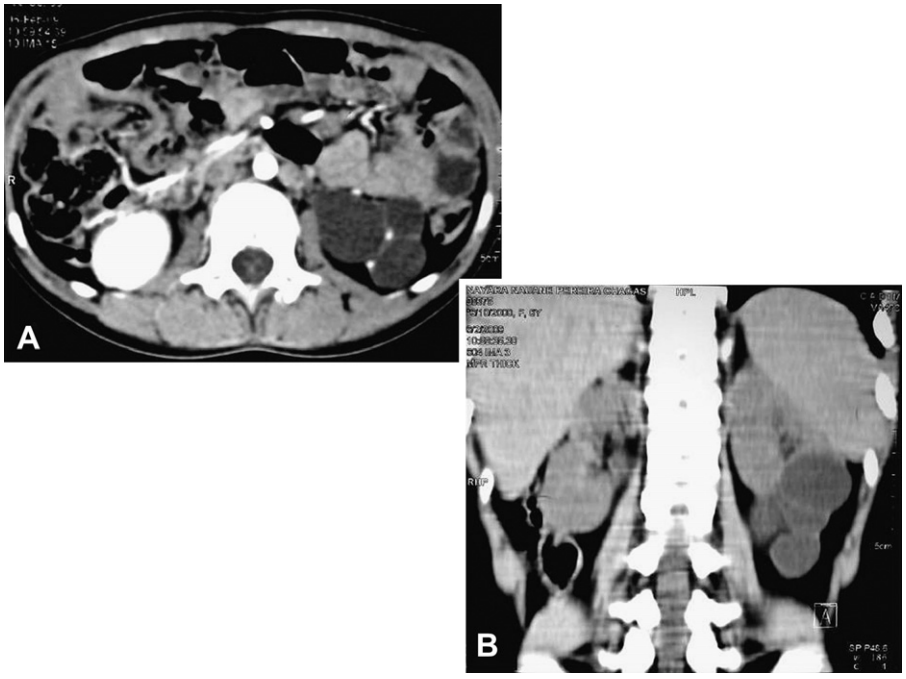

Figure 2 Non-contrast CT scan identifies clearly the cysts on the left lower pole.