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CASE REPORT

Unusual location and orientation of crossed fused kidneys in a patient with history of repaired congenital diaphragmatic hernia and hypoplastic lung: Importance of [^{99m}Tc]Tc-DMSA SPECT/CT and correlation with diuretic renal scan and VCUG

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ABSTRACT

We presented a 2.5-year-old boy with a history of the left pulmonary hypoplasia, repaired congenital diaphragmatic hernia and multiple episodes of pyelonephritis was referred to our department for [^{99m}Tc]Tc-DMSA and [^{99m}Tc]Tc-EC renal scintigraphy. The scans revealed an unusual pattern of renal uptake with horizontally aligned kidneys located high on left lower thoracic region extended to the mid abdomen. SPECT/CT of the [^{99m}Tc]Tc-DMSA scan clearly demonstrated the unusual location of the kidneys which was due to crossed fused renal ectopia and elevated diaphragm. Diuretic renal scans showed no evidence of renal obstruction. Additionally, the VCUG scan indicated vesicoureteral reflux in both ureters.

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CASE PRESENTATION

A 2.5-year-old boy with history of the left pulmonary hypoplasia, repaired congenital diaphragmatic hernia and multiple episodes of pyelonephritis was referred to our nuclear medicine center for [^{99m}Tc]Tc-DMSA renal scintigraphy due to fever and acute pyelonephritis. The planar [^{99m}Tc]Tc-DMSA images (Figure 1A) showed unusual shape and location of fused kidneys with horizontal orientation on the left side of the body.

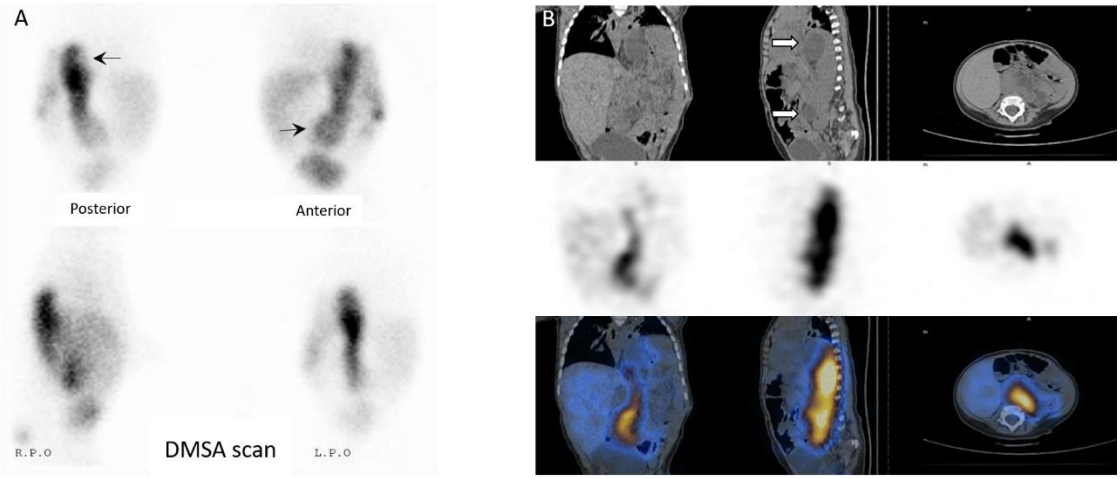

Figure 1. Planar (A) and SPECT/CT (B) [^{99m}Tc]Tc-DMSA scan of the patient. Arrows denote dilated PC systems

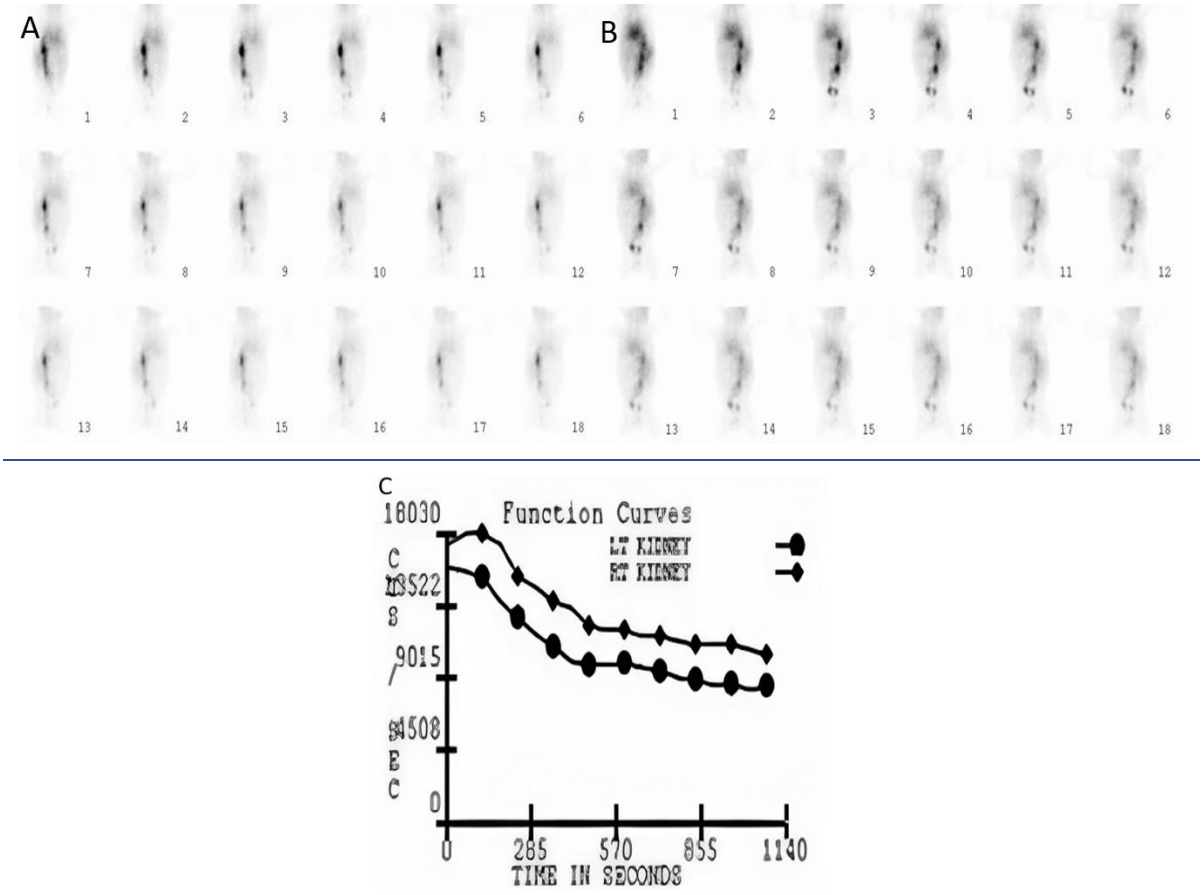

Figure 2. [^{99m}Tc]Tc-EC scan of the patient in posterior (A), and anterior (B) views. Note the renogram on the right side of the figure (C)

Two areas of decreased uptake was suspicious of being cortical defects (arrows). The patient underwent SPECT/CT imaging (Figure 1B) which revealed a single kidney with homogenous [^{99m}Tc]Tc-DMSA uptake on the left lower thoracic region extending to the mid abdomen. Areas of decreased activity were proven to be dilated pyelocalyceal systems (arrows). The kidney was abutting the left diaphragm, which was elevated due to left lung hypoplasia (Figure 1). The [^{99m}Tc]Tc-EC scan revealed fused kidneys with two ureters on the left side of the body and good response to lasix injection. Dynamic images showed two pyelocaliceal systems, and the time-activity curve (of the posterior view) (C) indicated normal initial uptake without evidence of obstruction (Figure 2). Additionally, the voiding cystourethrogram (VCUG) scan showed vesicoureteral reflux in both ureters (arrows) (Figure 3).

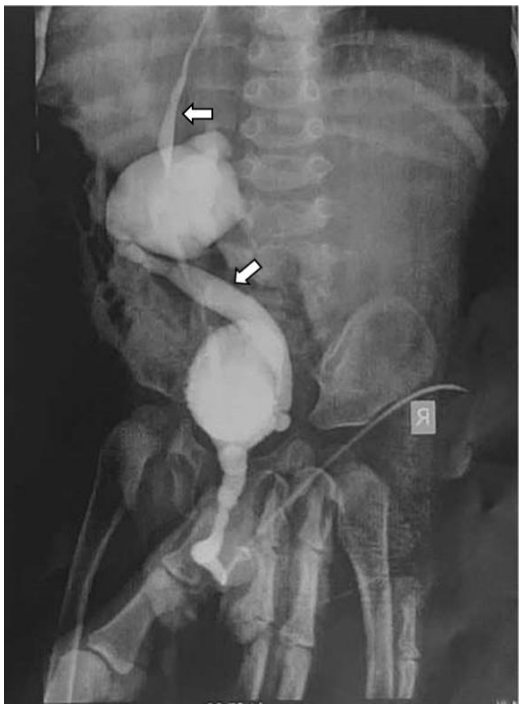

Figure 3. VCUG scan of the patient. Arrows denote reflux to the pelvicalyceal (PC) systems

DISCUSSION

Congenital kidney and urinary tract anomalies are commonly diagnosed in newborns. Crossed fused renal ectopia (CFRE) is a rare congenital urinary tract malformation related to migration defect and is considered as the second most common renal fusion anomaly after horseshoe kidney. In this anomaly, the kidney is located on one side, with its ureter entering the bladder on the opposite side [1, 2]. The true incidence of CFRE is not yet known but has been estimated to be around 1:2000 to 1:7500 autopsies [3]. There have been six reported variations of crossed fusion in the kidneys: type 1, where the kidneys are fused in an inferior position; type 2, where they form a sigmoid or S-shape; type 3, a unilateral lump kidney; type 4, a unilateral disc kidney; type 5, an L-shaped kidney; and type 6, where the fusion occurs in a superior position. Based on this classification system, the patient in question was diagnosed with type 6 crossed fused renal ectopia [4]. Imaging studies revealed type 2 crossed fusion in our patients. This anomaly often remains asymptomatic and is incidentally discovered during screening for associated congenital anomalies or evaluation for other conditions [5]. In some cases, it may be associated with recurrent urinary tract infections, vesicoureteral reflux, nephrolithiasis, hydronephrosis, ureteropelvic junction obstruction, and multicystic renal dysplasia [3, 5, 6].

Further imaging studies are usually required in case of suspicious or proven CFRE. The [^{99m}Tc]Tc-DMSA scan can reveal the location of functioning renal tissue with a high degree of sensitivity [7-9], and additional SPECT/CT imaging may be helpful in identifying any morphologic anomalies and cortical defects [10]. Diuretic renal scan would also be useful in identifying renal outflow obstruction and estimating its impact on renal function [9-12].

CONCLUSION

Nuclear medicine can be helpful in determining the type of anomaly and detecting possible cortical defects or obstruction. Our study highlights the importance of SPECT/CT images for correct diagnosis of renal anomalies and normal variations.

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