Case Report

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TANAFFOS

Ectopic Intrathoracic Kidney Associated with Ipsilateral Ectopic Spleen and Diaphragmatic Hernia in a Pediatric Patient: A Case Report

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Background: Intrathoracic kidney is the rarest form of an ectopic kidney that is usually accompanied by left congenital diaphragmatic hernia (CDH) (Bochdalek hernia), the association of which with other anomalies is rare.

Case Presentation: Herein, we describe a case with a diagnosis of an intrathoracic kidney associated with the ectopic spleen and diaphragmatic hernia diagnosed during imaging studies for urinary tract infections (UTIs). This study reports an 11-month-old male case with a history of CDH and several episodes of UTIs. A kidney ultrasound revealed that the left kidney and spleen were located in the thoracic cavity. Despite intrathoracic lying of the left kidney, there was no vesicoureteral reflux. Technetium-99m dimercaptosuccinic acid scan reported a highly positioned left kidney .

Conclusion: With the consideration of a pediatric literature review among patients with intrathoracic ectopic kidney, our case was special and notable since it was the first neonate who had an association of intrathoracic spleen and kidney in the same side with a delayed diagnosis. The main point of this case was that radiologists should consider thoracic kidney a differential diagnosis of unilateral renal agenesis when there is a history of diaphragmatic hernia.

Key words: Intrathoracic kidney, Ectopic kidney, Ectopic spleen, Diaphragmatic hernia

INTRODUCTION

In 1848, Vincenz Alexander Bochdalek first discovered posterolateral congenital diaphragmatic hernia (CDH) (1). Despite the low estimated incidence of this neonatal abnormality (1 in 13,000), it can bring high morbidity and mortality rate (up to 80%) (2-4). The left side is the most desired place for these hernias to occur (80-90%) (5). Although CHD can be accompanied by ectopic thoracic kidneys, they are still rare entities, approximately 5% of all renal ectopia, usually asymptomatic, most notably in adults (6). In children, most cases are recognized prenatally or after delivery; however, 5-25% of them can be presented lately with gastrointestinal problems, cardiopulmonary impairment, and urinary tract disorders (7, 8).

Among live-born infants with CDH, 40-60% of them have associated anomalies; however, the combination of kidney and spleen ectopia has not been reported in children and, to the best of our knowledge, only in an adolescent patient (4, 9, 10). Herein, we report a rare case of ipsilateral double ectopia (intrathoracic kidney and spleen) due to a left Bochdalek hernia with an unusual presentation in infancy.

CASE SUMMARIES

An 11-month-old male infant was referred due to multiple hospital admissions for urinary tract infections (UTIs). There was a history of surgical operation to repair a congenital Bochdalek-type diaphragmatic hernia in the neonatal period. At the age of 50 days, following the episode of breathlessness, fever, nausea, vomiting, and diarrhea, with positive urine culture for *Klebsiella pneumoniae*, the diagnosis of UTI was made for the patient. After a couple of months, he was admitted to an intensive care unit due to sepsis evaluation with positive urine and blood culture for *Klebsiella pneumoniae*. At that time, he had a normal abdominal ultrasound, chest x-ray, and normal voiding cystourethrogram, and his physical examination was normal. Technetium-99m dimercaptosuccinic acid (Tc-99m DMSA) scan was performed due to repeated pyelonephritis in infancy (Figure 1). A highly positioned left kidney was noted that was not emphasized by the radiologist .

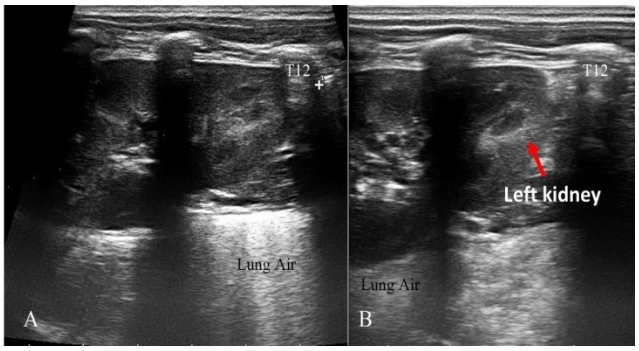

Figure 1. The posterior sagittal view of the intrathoracic kidney: the lower pole of the left kidney is above the latest rib in expiration time (A), and at its level during inspiration (B). The kidney is located between the thoracic wall and the air in the lungs.

Prophylactic antibiotics were recommended to prevent recurrent UTIs. At the age of 18 months, due to the repeated episodes of diarrhea accompanied by irritability, an abdominal ultrasound was requested to rule out intussusception. At that time, physical examination was normal. The left kidney was not detected, and the radiologist checked the thoracic cavity due to the absence of the spleen in the abdominal cavity. It revealed double ectopia in the left side, including kidney and spleen, probably herniated through the left posterolateral diaphragmatic defect into the left hemithorax (Figure 2). There was no abnormal renal rotation.

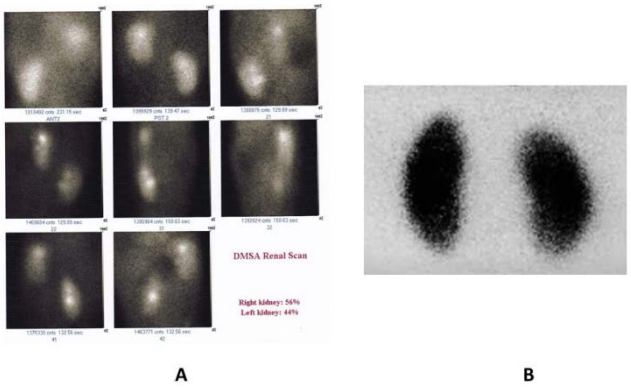

Figure 2. Tc99m-DMSA scan demonstrated normal findings. No defect was reported, however; the left kidney was seen in higher position than the right one (A) compared with normal TC99 DMSA scan (B) .Both kidneys had normal uptake with differential renal function 56% and 44% in right and left kidney, respectively.

DISCUSSION

The Bochdalek-type of CDH tends to occur in the left side (80-90%), since the early closure of right foramen in fetal life than the left one (11, 12). The association of CDH with intrathoracic renal ectopia has been observed with the incidence of 1 in 16,000 cases (11, 13). Our case is a male infant with a late diagnosis of double left ectopia (thoracic kidney and spleen) with a left Bochdalek hernia following vague findings. Intrathoracic ectopic kidneys can be associated with a combination of gastrointestinal or urinary anomalies; however, our case was unique since the combination of spleen and kidney ectopia was scarce (14).

An intrathoracic ectopic kidney has various ranges of differential diagnosis, such as esophageal or bronchogenic cyst, pulmonary sequestration, and even aneurysm of the descending aorta; nevertheless, it usually presents with gastrointestinal and respiratory symptoms (15). Our case suffered from recurrent UTIs and was initially missed at the referred center. The confirmed diagnosis of this anomaly is based on computed tomography (CT) scan; however, it is acceptable that the early

diagnosis in neonates can be possible with the aid of ultrasound (16, 17).

In the past, the modality of choice for confirming the diagnosis of an intrathoracic kidney was intravenous urography. Recently, an ultrasound and a CT scan are the choice modalities (18, 19). Additionally, Tc-99m DMSA scan and Tc-99m diethylenetriaminepentaacetic acid scintigraphy can differentiate an ectopic thoracic kidney from other tissues. Tc-99m DMSA scan is preferred in case of the need for the assessment of renal scarring, absolute divided renal function, or functioning of renal parenchyma (20).

The present case is special due to the rare association of thoracic ectopic spleen with ipsilateral thoracic kidney. This case highlights the consideration of the ectopic thoracic kidney in the differential diagnosis of a child with urinary infection when one kidney is not detected in the abdominal cavity. In our case, if the chest ultrasound was not performed, it could be considered unilateral renal agenesis.

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