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Thoracic Ectopic Kidney with Diaphragmatic Hernia

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ABSTRACT

Congenital thoracic ectopic kidney is a very rare developmental anomaly and the rarest form of all ectopic kidneys .it is usually asymptomatic and discovered incidentally in routine chest radiography.

We report an 18-month old boy with right congenital diaphragmatic hernia with thoracic ectopic kidney and positional respiratory symptoms.

Chest X- ray revealed opacity at the base of right lung. Dimercaptosuccinic acid (DMSA) scan showed right thoracic kidney.

Hereby we discuss the features of congenital right thoracic ectopic kidney and review the literature in this regard. (Tanaffos 2006; 5(2): 69-72)

Key words: *Ectopic kidney, Diaphragmatic hernia, Dimercaptosuccinic acid (DMSA), Positional respiratory symptoms.*

INTRODUCTION

Thoracic kidney is a very rare form of renal ectopia exists when the kidney is positioned considerably higher than normal. Intrathoracic ectopia denotes either a partial or a complete protrusion of kidney above the level of the diaphragm into the posterior mediastinum. The incidence of ectopic kidney is 1/1000 birth but less than 10% are detected (1, 2). Fewer than 5% of all patients with renal ectopia have an intrathoracic kidney (1, 3).

Since 1988, at least 140 cases with thoracic kidney have been reported in the literature, 4 of whom had bilateral thoracic kidney. (3) There appears to be a slight left-sided predominancy 1.5:1, and the sex ratio favor male by 2:1(4, 5, 6).

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Embryology: The kidney reaches its adult location by the end of the 8th week of gestation. At this time, the diaphragm leaflets are formed as the pleuraperitoneal membrane separate the pleura from the peritoneal cavity (7).

Mesenchymal tissues along with this membrane eventually form the muscular component of the diaphragm. It is uncertain whether delayed closure of diaphragmatic anlagen allows for protracted renal ascent above the level of the future diaphragm, or the kidney overshoots its usual position because of the accelerated ascent before normal diaphragmatic closure (3, 7).

The kidney is situated in the posterior mediastinum and usually has completed the normal rotation process. Therefore, the kidney is not within the pleural space, and there is no pneumothorax (3, 8).

The lower lobe of the adjacent lung may be hypoplastic secondary to compression by the kidney mass (4).

The renal vasculature and the ureter enter and exit from the pleural cavity through the foramen of Bochdalek.

The ureter is elongated to accommodate the excessive distance to the bladder, but usually it never enters ectopically into the bladder or other pelvic sites. (3) No consistent anomalies have been described in other organ systems, except, one child who had trisomy -18 and another patient had multiple pulmonary and cardiac anomalies in addition to the thoracic kidney (2, 3).

The vast majority of affected individuals have been remained asymptomatic. Pulmonary symptoms are exceedingly rare, and urinary ones are even more infrequent (3).

Most cases are discovered on routine chest radiography or at the time of thorcotomy for a suspected mediastinal mass (1, 3).

The diagnosis is most commonly made after routine chest radiography in which the affected hemidiaphragm is found to be elevated slightly (3, 9).

Most patients are discovered fortuitously and have no specific symptoms referable to the misplaced kidney. No treatment is necessary once the diagnosis has been confirmed (4, 10).

CASE PRESENTATION

An 18-month old boy who was a known case of hypospadias referred to the clinic for further investigation. He had history of breathing difficulties on supine position since two weeks prior to our first visit.

His hypospadias was glandular type and located on glans of his penis which is the most common form. There was not any apparent other anomaly of external genitalia, but in his chest examination the breathing sound was decreased in lower part of right hemithorax.

Chest radiography showed an opacity and air blebs at the base of right lung. (Figure-1)



Figure 1. CXR of patient.

On abdominal sonography the right kidney was not seen in its normal location, but was located in right thoracic space and its size was at least 2cm smaller than the left side.

Further investigation yielded the following results

- Demercaptosuccinicacid (DMSA) Scan showed right thoracic kidney with proper function, and with no scar (Figure-2).

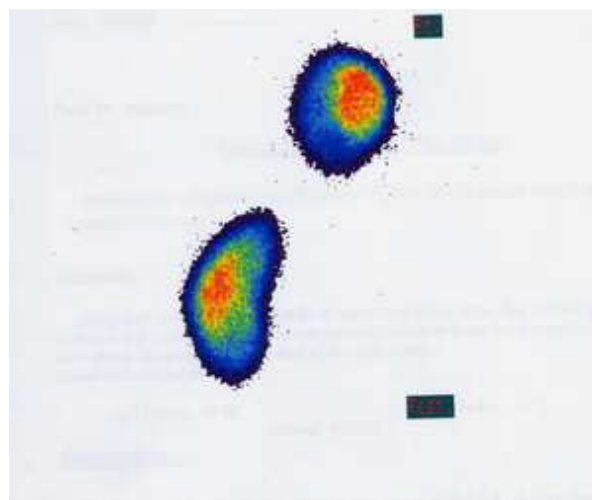


Figure 2. DMSA scan of patient

- Upper gastrointestinal (GI) series showed right diaphragmatic hernia with bowel displacement in thoracic cavity. (Figure-3)



Figure 3. Upper GI series of patient.

-Lower esophageal sonography showed gastroesophageal reflux with mucosal thickening.

- Magnetic resonance urography (MRU) revealed no anomalies except the thoracic kidney. (Figure-4)



Figure 4. Magnetic resonance urography of patient.

- Magnetic resonance angiography (MRA) showed the right artery arising from abdominal aorta. (Figure-5).



Figure 5. Magnetic resonance angiography of patient.

- Voiding cystourethrogram (VCUG) of patient showed grade two right vesicoureteral reflux (Figure-6)



Figure 6. Voiding cystourethrogram of patient.

Correction of diaphragmatic hernia was performed by pediatric surgeon.

DISCUSSION

We described an 18-month old boy with ectopic thoracic kidney along with diaphragmatic hernia. He was healthy since two weeks before our first visit, when his mother noticed a mild respiratory distress and coughing of her baby on supine position.

Usually the vast majority of patients who have ectopic thoracic kidney are asymptomatic and respiratory symptoms are exceedingly rare (3, 11, 12) but our patient had diaphragmatic hernia, thoracic kidney and positional respiratory symptoms. The patient had also vesicoureteral reflux in translocated kidney. On the other hand, the most common types of ectopic kidney and the most diaphragmatic hernia are located in the left side,(4, 5) but our patient had right sided thoracic ectopic kidney and right sided diaphragmatic hernia.

We recommend imaging investigation for infants and children with positional respiratory symptoms. We also emphasize that:

- a) Hypospadias may be associated with another significant anomaly of genitourinary or extra-genitourinary systems.
- b) Intrathoracic kidney and diaphragmatic hernia may present with positional respiratory distress or coughing.
- c) And last but not least, we recommend echocardiography because of cardiac anomalies in addition to the thoracic kidney (13).

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