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Short Commentary

Intrathoracic Renal Ectopia with Ipsilateral Congenital Diaphragmatic Hernia

Xin Sun, Dexin Yu^{*}

Department of Urology, The Second Hospital of Anhui Medical University, Hefei, Anhui, P.R. China.

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DESCRIPTION

The complex embryogenesis of the kidney may lead to several renal anomalies, renal ectopia included. Of all kidney ectopias, the intrathoracic one is the least common. Congenital diaphragmatic hernia is usually seen in the neonatal period, mainly because of failure to close the posterolateral diaphragm. Intrathoracic renal ectopia due to congenital diaphragmatic hernia has a reported incidence of 0.25% [1]. The association of a duplex kidney collection system and an intrathoracic renal ectopia is extremely rare. So far, only one infant patient case has been reported [2]. Herein, we report on an adult case of intrathoracic renal ectopia with a double kidney collection system associated with an Ipsilateral congenital diaphragmatic hernia.

A 51-year-old man presented to the urology clinic with a 3day history of slight backache after fatigue. He denied any history of trauma or hypertension, and his symptoms improved after resting. On physical examination, he only had slightly decreased left lower respiratory sounds. His blood pressure was 129/81 mm Hg. Urinalysis was without hematuria or proteinuria. Her serum hemoglobin was normal at 145 g/l and serum creatinine was 0.90 mg/dl. Other laboratory tests and pulmonary function tests also showed no obvious abnormalities. Chest, abdomen, and pelvis computed tomography showed that a left Intrathoracic renal ectopia with ipsilateral congenital diaphragmatic hernia. Intravenous pyelography performed in both the supine and erect positions revealed that his left kidney had duplex renal pelvis and ureters. The upper renal pelvis and ureter were located in the thoracic cavity, while the two ureters converged in the middle of the ureter (Fig. 1). Therefore, the patient was diagnosed with a left Intrathoracic renal ectopia with duplicate renal pelvis and ureter, accompanied by an ipsilateral congenital diaphragmatic hernia.

*Corresponding author. Dexin Yu, E-mail: dxyu@ahmu.edu.cn.

The patient's laboratory tests and pulmonary function tests were normal. No symptoms of severe backache, hematuria, or hydronephrosis were detected. Thus, he received conservative treatment. During the one-year follow-up, he recovered well, with no evidence of disease progression.



Figure 1: (a) Computed tomography imaging in the supine position of left intrathoracic ectopic kidney with diaphragmatic hernia. (b) Intravenous pyelography performed in the erect position demonstrating the location of the left kidney did not change significantly, which had duplicate renal pelvis and ureters.

DISCUSSION

Intrathoracic renal ectopia accompanied with congenital diaphragmatic hernia often was asymptomatic. Renal dysfunction is not common. Most patients are diagnosed accidentally or at autopsy, the treatment is not necessary unless severe urinary or pulmonary complication occurs [3]. The most important differential diagnosis of intrathoracic renal ectopia is nephroptosis. The definition of nephroptosis is a renal descent of more than two vertebral bodies when the patient moves from a supine to an erect position. Flank pain when standing upright that resolves on lying down is the most common symptom. Moreover, nephroptosis can be associated with hydronephrosis, impaired drainage, hematuria, renal calculi, and hypertension. Nonetheless, nephroptosis usually occur in young asthenic females, most of which are asymptomatic and do not require surgical intervention [4]. In our case, the location of the left kidney did not move significantly with the change of body positi on. The diagnosis of nephroptosis was not established.

Differential diagnoses also contain some congenital diseases caused by genetic mutations, such as craniofrontonasal syndrome or Kabuki syndrome [5, 6]. These diseases are manifested as anomalies of multiple organs, the urinary system included, combined with a congenital diaphragmatic hernia and usually required surgical treatment.

Declarations

Conflicts of interest the author declare that they have no conflict of interest.

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