Diagnosis of Unilateral Thoracic Kidney by Color Doppler Ultrasound

Musarrat Hasan1*, Erum Saba1, Amir Bhatti2, Beenish Kanwal1

The incidence of ectopic kidneys is one in 900 autopsies. Thoracic kidney is the rarest form of all renal ectopias and is mostly asymptomatic. Intrathoracic kidneys may be congenital or acquired. We present here the case of a 13-year-old boy who presented with the complaint of mild dyspnea even at rest, which had occurred periodically since birth. An X-ray showed obliterated costophrenic angles. He was referred to our department for ultrasound examination of the whole abdomen. Upon examination by ultrasound, both kidneys were not visualized in the lumbar region or in the lower abdomen. Therefore, color Doppler was used to locate the renal arteries. Both renal arteries were observed arising from their usual site and ascending upwards. The left kidney was visualized in the chest; it was malrotated and placed transversely above the diaphragm. The right kidney was visualized higher up in the chest but below the diaphragm. A computed tomography pyelogram and computed tomography angiogram also confirmed our findings. No intervention was required because the intrathoracic location of a kidney typically does not affect renal function and does not lead to other symptoms.

KEY WORDS — color Doppler ultrasound, computed tomography pyelogram, thoracic kidney


Introduction

Ectopic kidney occurs in one of 900 autopsies and its occurrence is similar in both sexes [1]. An ectopic kidney can be pelvic, iliac, abdominal, thoracic or crossed [1]. Thoracic kidney is the rarest form of all renal ectopias with a prevalence of less than one in 10,000 [2,3]. It is mostly asymptomatic and results from an abnormally high descent of the metanephros, which generates a diaphragmatic defect and subsequently an ectopic kidney in the thorax. It is uncertain whether delayed closure of the diaphragmatic analog allows for protracted renal ascent above the level of the future diaphragm, or whether the kidney overshoots its usual position because of accelerated ascent before normal diaphragmatic closure. The kidney usually lies in the posterolateral aspect of the diaphragm in the foramen of Bochdalek. The diaphragm thins out at this point, and a flimsy membrane surrounds the protruding portion.
of the kidney. Therefore, the kidney is not within the pleural space and there is no pneumothorax. The adrenal gland is below the kidney in its normal location in most patients and in unilateral cases the contralateral kidney is usually normal [4].

Intrathoracic kidneys may be congenital or acquired. This condition is rarely bilateral (10%), and when it is unilateral, it occurs most commonly on the left side. There is a marked preponderance in males with a ratio of 2:1 [5,6]. Intrathoracic kidneys are most often discovered as a posterior mediastinal or juxtadiaphragmatic mass by chest radiography. They can be further evaluated by intravenous urography or sonography, which allows differentiation from significant lesions, thus preventing unnecessary surgery or biopsy [3,7]. Prenatal diagnosis of intrathoracic kidneys by ultrasonography is possible [8], but they may be overlooked by routine prenatal examination. Perhaps surprisingly for nonsurgeons, ectopic intrathoracic kidney is not always associated with diaphragmatic hernia [9]. Most patients with ectopic thoracic kidney have a normal diaphragm with an intrathoracic but extrapleurally located kidney (true ectopic kidney). Ectopic thoracic kidney may also result from eventration of the diaphragm (relaxation diaphragmatica), diaphragmatic hernia, or traumatic rupture of the diaphragm [10]. We report here the case of a boy with intrathoracic kidneys diagnosed using color Doppler ultrasound.

**Case Report**

A 13-year-old boy presented with the complaint of mild dyspnea that had occurred periodically since birth. An X-ray showed obliterated costophrenic angles with an increased anteroposterior diameter of the thoracic cage. There was also evidence of elevation of both hemidiaphragms, which was more marked on the right side. The lung fields showed no abnormality.

The patient was referred to us for ultrasound examination of the whole abdomen. Upon examination by ultrasound, both kidneys were not visualized in the lumbar region or in the lower abdomen. Therefore, color Doppler was used to locate the renal arteries. Both renal arteries were seen arising from their usual site and ascending upwards (Fig. 1). Ultrasound was carried out in the intercostal region at the level of the pleural space and the left kidney was visualized in the chest because the diaphragm was clearly visualized. The left kidney was malrotated and was placed transversely above the diaphragm. The right kidney was visualized higher in the chest and placed very close to the spine (Fig. 2); it was difficult to determine whether it was above the diaphragm or whether it was just evagination of the diaphragm (due to weakening).

![Fig. 1. Both renal arteries were seen arising from their usual site and ascending upwards. RV = right vein; RA = right artery; L = liver.](image1)

![Fig. 2. The right kidney (RK) was visualized higher in the chest and placed very close to the spine. IVC = inferior vena cava; L = liver.](image2)
To confirm our diagnosis, we carried out an intravenous pyelogram, which showed normal functioning kidneys. The intravenous pyelogram showed that the right kidney was positioned higher up but that it remained below the right hemidiaphragm. The left kidney appeared to be above the left hemidiaphragm. The urinary bladder appeared normal with a smooth outline. No calculus in the kidneys, ureters or urinary bladder was noted.

A computed tomography (CT) pyelogram and CT angiogram showed elevation of the right hemidiaphragm (Figs. 3 and 4). The right kidney was observed in a high position yet it was below the diaphragm. It was also malrotated and placed medially with a very close approximation of its upper pole with the inferior vena cava. The left kidney was observed lying above the diaphragm (intrathoracic in location). It was rotated along its long axis and was observed lying in a transverse position with its pelvi-ureteric junction facing below. Both kidneys, however, showed normal function. No calculus or hydronephrosis was noted. Marked hepatosplenomegaly was also observed with a triangular segment of an enlarged liver, which seem to be an accessory segment of the liver. Malrotation of the liver was observed and an accessory spleen was noted. Herniated large bowel loops were observed below the right hemidiaphragm lateral to the right kidney.

**Discussion**

Congenital anomalies of the urinary system affect approximately 10% of the population [11]. Unilateral thoracic (or superior) renal ectopia is a rare anomaly occurring only once in a series of 13,000 autopsies and accounts for 5% of all renal ectopias [2,12]. The early signs of kidney development can be seen at the beginning of the 4th week of pregnancy. A ureteral bud separates from the Wolffian duct and ascends toward the urogenital ridge. In the 5th week, the metanephric blastema develops, appearing above the migrating bud. A period of rapid caudal growth in the embryo appears to assist in migration of this structure out of the pelvis and into its eventual retroperitoneal location in the renal fossa. By the 8th week, migration and rotation appear to be complete. Factors that interfere with development such as teratogens, genetic factors, ureteral buds or metanephric maternal disease may result in abnormal migration of the kidney and renal ectopia. The vascular supply to the organ changes several times during the migratory event, and abnormal origins of renal arteries are related to when migration is arrested [1,13].

Unlike pelvic ectopic kidneys, which may be obstructed, infected or have calculi, thoracic ectopic kidneys are usually normal [14,15]. They can, very rarely, be affected by a lung abscess resulting in
sputum with a foul urine smell [16], or a renal cell carcinoma may develop in an intrathoracic kidney [17]. Thoracic ectopic kidneys may be discovered by chance during complementary exploration of acute respiratory tract pathology or a lung mass [18–21]. With advances in the field of ultrasonography, prenatal diagnosis of intrathoracic kidneys has been made using color Doppler ultrasound [8].

Except for a few cases in which thoracic ectopic kidney was found after traumatic diaphragmatic rupture [22,23], most thoracic ectopic kidneys are assumed to be congenital. In some cases, superior origin of the renal vessels has been noted; however, in a few other cases it was simply longer than normal [8]. Occasionally the kidney resides entirely above the diaphragm, but most commonly it occupies a posteromedial diaphragmatic defect that may or may not be capped by fibrous tissue as observed by surgery or autopsy [23,24]. Superior migration of the metanephros before completion of diaphragmatic development during the 8th week of gestation is common in most embryological theories [24–26]. Pfister-Goedek and Brunner [27] have classified the anomalies into four entities: thoracic renal ectopia with a closed diaphragm, thoracic renal ectopia with entervation of the diaphragm, thoracic renal ectopia with diaphragmatic hernia (congenital and acquired), and traumatic rupture of the diaphragm with renal ectopia.

An empty renal fossa should not be overlooked. Doppler ultrasound of the aorta can be used to reach the origin of the renal vessels. In our study, Doppler ultrasound of the renal vessels was carried out and a sample was obtained from the right renal artery, which was observed ascending towards the diaphragm. The left renal artery was observed on color Doppler ultrasound but it was difficult to obtain a spectral sample because of gas shadows. Ultrasound was then carried out in the intercostal region at the level of the pleural space and the left kidney was found lying in the chest above the diaphragm. The left kidney was malrotated and was placed transversely above the diaphragm. The right kidney was visualized higher up in the chest and was placed very close to the spine.

True intrathoracic ectopic kidney presents during fetal life and has four characteristics: (1) a rotation anomaly, (2) a long ureter, (3) anomalous high derivation of the renal vessels from the thoracic aorta, and (4) medial deviation of the lower pole of the kidney [5]. A few cases of thoracic kidney accompanied by superior ectopic spleen have been published [11,28]. Thoracic kidney, although extremely rare, must always be considered in patients with an intrathoracic mass on a chest radiograph, even if this was not present in a previous examination [29,30]. Ultrasonography, CT pyelography and CT angiography may be helpful in the identification of this anomaly. Because the intrathoracic location of a kidney typically does not affect renal function or cause symptoms, it requires no intervention [14,15].

References