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Bilateral thoracic kidneys combined with inferior vena cava located behind the anterior abdominal wall: A case report and review of literature

Xiao-Xia Peng, Shao-Ang Cheng, Qi-Lian Liang, Xing-Bo Luo, Xiao-Cui Hong, Gao-Le Yuan, Hui-Jie Zhang

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Abstract

Ectopic thoracic kidneys are the rarest form of renal ectopia. Moreover, congenital abnormality of a primary anterior inferior vena cava (IVC) located behind the anterior abdominal wall is extremely rare. To date, only one such case has been reported. Herein, we report a rare case of a 55-year-old Chinese male with bilateral thoracic kidneys combined with an anterior IVC, a malformed liver, and a large-round-folds navel. The classification, clinical characteristics, and management options of a thoracic kidney was also summarized by literature review. To our best knowledge, the simultaneous detection of such multiple complex abnormalities has not been reported.

Key words: Thoracic/intrathoracic kidney; Inferior vena cava; Ectopic kidney; Renal ectopia; Congenital anomaly

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Core tip: We described an unusual case of bilateral thoracic kidneys combined with anomalies of the inferior vena cava, liver, and navel in this article. The classification, clinical characteristics, and management

options of a thoracic kidney was also summarized by literature review.

Peng XX, Cheng SA, Liang QL, Luo XB, Hong XC, Yuan GL, Zhang HJ. Bilateral thoracic kidneys combined with inferior vena cava located behind the anterior abdominal wall: A case report and review of literature. *World J Clin Cases* 2018; 6(13): 666-670 Available from: URL: <http://www.wjgnet.com/2307-8960/full/v6/i13/666.htm> DOI: <http://dx.doi.org/10.12998/wjcc.v6.i13.666>

INTRODUCTION

Renal ectopia is an anatomic anomaly where the kidney is located outside the normal renal fossa. The prevalence of this condition is about 1 in 1000 live births, but only about 10% are diagnosed^[1]. Thoracic renal ectopia refers to the partial or complete protrusion of the kidney into the thorax. In contrast to ectopic kidney in the pelvic, lumbar, and iliac areas, the ectopic thoracic kidney is the rarest form of anomaly with an incidence rate of less than 5% of the renal ectopias and can be discovered only due to discomfort or incidental detection by chest radiography^[2,3]. Only six cases of bilateral thoracic kidneys have been reported in the literature^[4,5]. An intrathoracic kidney usually remains asymptomatic throughout a patient's life and has a good prognosis without any intervention therapy.

The inferior vena cava (IVC) is the largest retro-peritoneal vessel responsible for transporting most venous blood from the abdomen and lower extremities back to the right atrium of the heart^[6]. Normal IVC originates from the confluence of the right and left common iliac veins at vertebral level L5, lies along the right anterolateral aspect of the vertebral column, passes through the central tendon of the diaphragm around vertebral level T8 and enters the posterior inferior aspect of the right atrium. Commonly, the normal IVC is composed of four segments: hepatic, suprarenal, renal, and infrarenal. It is developed from the vitelline veins and the three paired cardinal veins including subcardinal veins, sacrocardinal veins, and the supracardinal veins during embryonic development^[7]. These venous systems underwent a complex procedure involving development, regression, anastomosis, and replacement in the formation of IVC^[8]. The failure of any process in the formation procedure of IVC can result in anatomic abnormalities of the IVC.

Congenital abnormalities of the IVC are complex and variable; they have become increasingly recognized given the increased use of cross-sectional imaging. Many reports of IVC anomalies, including a left IVC, double IVC, intrahepatic IVC agenesis, and absent infrarenal IVC, have emerged. However, only one case of the congenital abnormality of a primary anterior IVC located behind the anterior abdominal wall was reported in 2011.

Although six cases of bilateral thoracic kidneys

have been documented in literature, none of them have been associated with an anterior IVC anomaly. In this text, we present the first case of simultaneous detection of bilateral thoracic kidneys and an anterior IVC, a malformed liver, and a large-round-folds navel. The anatomic abnormality described in our case was demonstrated by contrast-enhanced computed tomography (CT) and a technology of multiple dimensional reconstructions. We also summarized the clinical characteristics and management options of thoracic kidney by literature review.

CASE REPORT

A 55-year-old Chinese male presented with a complaint of progressively worsening intermittent abdominal blunt pain in the right lower quadrant for 2 mo without radiation pain. He denied nausea, vomiting, bloating, diarrhea, hematochezia, fever, chills, chest tightness, palpitations, difficulty breathing, and any urinary symptom or other discomforts. The patient underwent a colonoscopy scan a week prior at the outpatient clinic of our hospital that revealed an ileocecal mass confirmed as adenocarcinoma by histopathologic examination. No loss of appetite or weight was noted. Medical and surgical histories, as well as family history, were unremarkable. No past history suggestive of pulmonary disease or major trauma was elicited.

Physical examination revealed a well-developed thin man with several soft round skin folds approximately 10 cm in size at the position of the navel. In addition, tenderness was detected at the lower right abdominal quadrant. Edema of the lower extremities, varicosity, and other special findings were not found. Laboratory results showed a carbohydrate antigen 125 level of 38.42 U/mL (reference range, < 35.0) and a carbohydrate antigen 19-9 level of 773.3 U/mL (reference range, < 27.0). The values for routine blood chemistry and liver and kidney function examination were within normal limits.

CT scan was performed to evaluate for neoplastic infiltration. Unexpectedly, CT imaging located the double kidneys at an unusually superior position immediately below the diaphragm, with part of the kidney protruding into the thorax (Figure 1). A round, homogeneous, low-density cyst about 1.8 cm in size was depicted in the right kidney. The diaphragm was intact without other abdominal organs herniating into the chest cavity. The suprarenal segment of the IVC coursed cranially and anteriorly along the space between the liver and diaphragm (Figure 2A and 2B) and bridged the renal segment and the hepatic segment. The hepatic IVC was located posterior to the anterior abdominal wall (Figure 2C) and curved posteriorly to join the right atrium by traversing through the sternal part of diaphragm (Figure 2A). Local hepatic IVC stenosis (Figure 2A) was observed. The morphology of the liver was abnormal (Figure 3), and hepatic veins traverse and drained directly

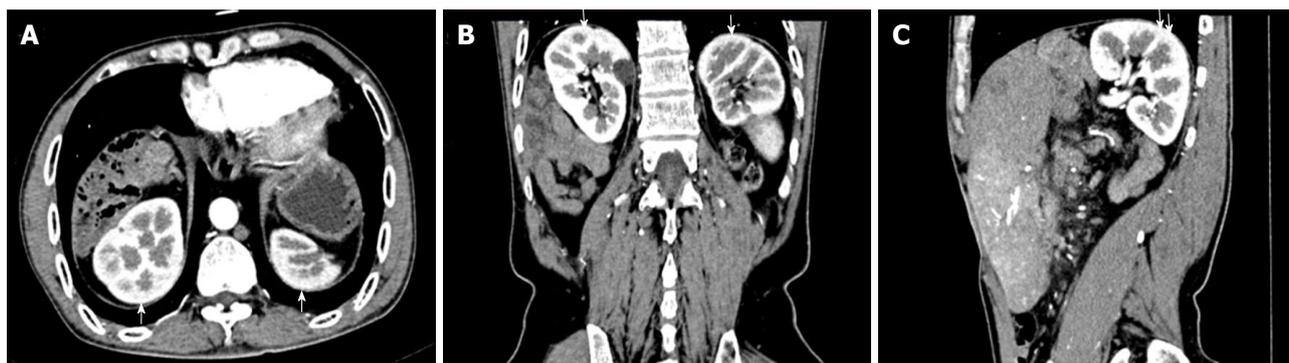


Figure 1 Contrast-enhanced computed tomography in arterial phase demonstrates the thoracic kidneys. A: Transverse view scan shows the upper pole of the kidney (arrows), above the liver, is surrounded by the lower lobe of lung; B: Coronal view scan shows the kidneys (arrows) located immediately below the diaphragm, with part of the kidney protruding into the thorax; C: Sagittal view scan shows kidney (arrows) located immediately below the diaphragm.

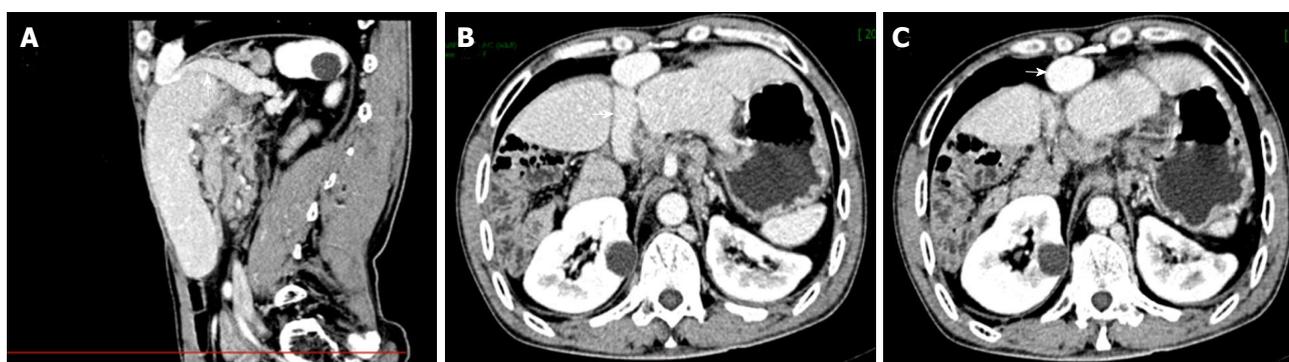


Figure 2 Contrast-enhanced computed tomography in venous phase shows heterotopic inferior vena cava located behind the anterior abdominal wall. A: Sagittal view scan demonstrates the suprarenal segment of the inferior vena cava (IVC) coursed cranially and anteriorly along the space between the liver and diaphragm and bridged hepatic segment and the renal segment; B: Transverse view scan shows the suprarenal segment of the IVC crossed through the liver and drained into the anterior IVC; C: Transverse view scan shows the hepatic IVC (arrow) was located posterior to the anterior abdominal wall.



Figure 3 Contrast-enhanced computed tomography in venous phase from cranial to caudal positions shows a liver with abnormal morphology. A: Transverse view scan above the hilar level of kidney shows the malformed liver (arrows); B: Transverse view scan at the hilar level of kidney shows the malformed liver (arrows); C: Transverse view scan below the hilar level of kidney shows the malformed liver (arrows).

into the anterior hepatic IVC. Also, contrast-enhanced CT showed a 2.4 cm × 3.0 cm × 2.3 cm, irregular, ill-defined mass at the right side of the pelvis, and a necrotic area was observed in the mass. Multiple enlarged lymph nodes were noted in the pelvic, retroperitoneal, and mesenteric areas.

In accordance with the patient's condition, we suggested that the patient be treated with neoadjuvant chemotherapy initially. However, the patient refused chemotherapy for various reasons and eventually underwent tumor resection by laparotomy. He was

discharged without further treatment after surgery. Six months later, the patient returned to the hospital because of abdominal pain and edema of the right lower limb and scrotum. We considered that the abdominal pain and edema were caused by the recurrence and the progression of the tumor.

DISCUSSION

Thoracic kidney is the rarest kind of renal ectopias, with only about 200 cases reported in the existing literature.

Campbell *et al*^[9] found only one thoracic kidney among 22 cases of ectopic kidneys in 15919 autopsies of children. Renal thoracic ectopia is more common at the left side than on the right side because the right hemidiaphragm anastomoses earlier on the right and the liver acts as a barrier. Such ectopia is also more common in males than females. In 1988, Donat *et al*^[10] reviewed 178 cases of intrathoracic kidney reported in literature from 1922 to 1986; the scholars found 61% of these cases lying on the left side, 36% on the right side, and 2% bilateral. About 63% of the thoracic kidneys were found in males and 37% in females. Our patient is the seventh report of bilateral thoracic kidneys.

Thoracic kidneys have been divided into four basic types: (1) total thoracic ectopia with intact diaphragm; (2) protruding from the diaphragm (also known as subdiaphragmatic kidneys); (3) congenital hernia (such as Bochdalek hernia); and (4) hernia after traumatic rupture of the diaphragm^[11]. Our patient's unremarkable history of surgery and trauma, as well as the intact diaphragm confirmed by coronal and sagittal CT images, is suggestive of diaphragm eventration, which belongs under the second type of thoracic kidney.

Although the majority of intrathoracic renal ectopias are asymptomatic and usually discovered incidentally during routine medical checks, a small number of sufferers may present with expiratory dyspnea, chest pain, polypnea, cough, and expectoration. Most cases manifest as an isolated abnormality, but concurrent aberrances have been reported in scattered literature. Such aberrances included genitourinary anomalies (such as ureteropelvic junction obstruction and ureteral duplication)^[12], wandering spleen, and cardiovascular abnormality (such as dextrocardia and patent ductus arteriosus). Jhun *et al*^[13] described a right thoracic kidney with a pulmonary vascular malformation in an adult. The patient we presented had bilateral thoracic kidneys with an anterior IVC and accompanying malformed liver and a large-round-folds navel, all of which has not been previously reported at the same time.

The anatomical features of ectopic thoracic kidneys are abnormal rotation, elongated ureters, higher origin, and longer length of renal arteries and veins compared to normal kidneys, and medial deviation of the inferior pole of the kidney. The location of the adrenal gland may be normal or also thoracic. In our case, the kidneys were normally rotated with a normal renal vessel anatomy and absent adrenal glands.

Upon literature review, we noted that the etiology of thoracic ectopic kidney combined with an unusual anterior IVC has not been described in the past. In the 6th and 9th weeks of embryonic development, the kidneys migrate cranially to their normal position. Failure in this process may result in renal ectopia. Some authors proposed that the late differentiation of metanephric tissue caused by delayed anastomosis between the ureteric bud and the metanephron leads to excessive renal ascent, which allows the kidney to overshoot from its normal position^[14]. Other scholars have postulated that an initial high cephalic origin of the renal germ

results in congenital thoracic kidney^[15]. An unusual case of kidney ascent in postnatal life has also been reported by Zolotas *et al*^[16] in 2016. In this case, a male infant's right kidney migrated gradually from the renal fossa to a higher position superior to the liver and below the diaphragm. Abdullah *et al*^[17] proposed that the anterior IVC may represent a failure of the evolution of the normal developmental anastomosis between the right subcardinal vein and the hepatic segment.

In 2003, Gayer *et al*^[18] suggested the potential association of IVC anomalies with renal aplasia. In 2016, Parmar *et al*^[5] reported the first case of a patient with ventral hernia after an omphalocele. The patient was finally diagnosed with bilateral subdiaphragmatic kidneys accompanied by spina bifida, retroaortic left renal vein, and azygos continuation of the IVC. However, whether the observation of concomitant bilateral thoracic kidneys with an unusual anterior location of the IVC and a large-round-folds navel in our case was an incidental finding remains to be determined. No research has suggested any association of a thoracic kidney with a heterotopic IVC located behind the anterior abdominal wall.

The combinations of various imaging techniques helps diagnose and differentiate the thoracic kidney from other thoracic masses and reduces the rate of missed diagnoses and misdiagnosis. This strategy is also helpful for evaluating renal function. Most thoracic ectopic kidneys are initially treated after the accidental discovery of abnormal masses on chest radiographs^[19]. Ultrasonography shows a high specificity for ectopic kidneys but occasionally misses the thoracic kidney. Renal scintigraphy is typically used to evaluate kidney function. A CT scan can delineate the aberrant position of the kidney. Magnetic resonance imaging alone confirms the relation to the diaphragm. Avni *et al*^[20] found that magnetic resonance imaging is better than ultrasonography for diagnosing in children. The thoracic kidney should constantly be considered as a differential diagnosis with diaphragmatic hernia, mediastinal lymphadenopathy, lung cancer, aortic aneurysm, and mediastinal tumor. Early diagnosis of the condition avoids unnecessary interventional examinations, pathological biopsy, and exploratory thoracotomy.

The treatment options for the thoracic kidney are not immutable. Generally, most people accept that neither medical nor surgical treatment is required for asymptomatic patients. Surgical intervention therapy for thoracic renal ectopia is suggested only in severe urinary symptoms or expiratory dyspnea and pulmonary infection determined to be caused by the thoracic kidney. Robotic laparoscopy has been successfully used in thoracorenal renal fixation many times and can be regarded as a replacement choice for laparotomy. Zolotas *et al*^[16] suggested that the thoracic kidney associated with Bochdalek hernia should undergo surgery to avoid splanchnocele formation and strangulation. In our case, the aberrant kidneys did not impair renal function and further interventions were not required.

In conclusion, an unusual case of bilateral thoracic kidneys combined with anomalies of the IVC, liver, and

navel was described in this article. Although the incidence of thoracic kidneys is low, clinicians and imaging doctors should consider this condition as a differential diagnosis for an abnormal lesion in the thorax. The majority of thorax kidneys have a good prognosis, and intervention therapy is not always necessary.

ARTICLE HIGHLIGHTS

Case characteristics

The patient accidentally discovered anatomic abnormalities due to the abdominal pain caused by bowel cancer.

Clinical diagnosis

The patient was diagnosed with: (1) colon cancer; (2) bilateral thoracic kidneys; (3) anterior inferior vena cava (IVC); and (4) liver malformation.

Differential diagnosis

Thoracic kidney, which showed as an abnormal shadow on the chest film, may usually be misdiagnosed as a diaphragmatic hernia, mediastinal lymphadenopathy, lung cancer, aortic aneurysm, and mediastinal tumor.

Imaging diagnosis

Contrast-enhanced computed tomography shows bilateral thoracic kidneys combined with an anterior IVC and a malformed liver.

Treatment

We advised regular follow-up for the congenital abnormalities of thoracic kidneys in this case.

Related reports

The IVC develops from the vitelline veins and the three paired cardinal veins including subcardinal veins, sacrocardinal veins and the supracardinal veins during embryonic development. The failure of the complex procedure involving development, regression, anastomosis, and replacement in the formation of IVC can result in anatomic abnormalities.

Term explanation

Thoracic kidney refers to the partial or complete protrusion of the kidney into the thorax. Anterior IVC refers to IVC located immediately behind the anterior abdominal wall.

Experiences and lessons

Though with low incidence, thoracic kidneys should always be taken into consideration as one of differential diagnosis for an abnormal lesion in the thorax and intervention therapy is not always necessary.

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