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Cephalad-renal ectopia: Bilateral subdiaphragmatic kidneys in a patient of omphalocele with ventral hernia

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Abstract

Renal ectopia is a rare congenital anomaly. Thoracic ectopic kidney was being considered as rarest, however no case of bilateral subdiaphragmatic kidneys in omphalocele patients presented with ventral hernia has been reported yet, as per our best of knowledge. This is a report of a 5-year-old male patient who presented with ventral hernia after omphalocele. A thorough examination, laboratory, and radiological investigations including ultrasonography, plain abdominal x-ray, intravenous urogram, and computerized tomography revealed bilateral subdiaphragmatic ectopic kidneys with azygos continuation of inferior vena cava, retro-aortic left renal vein and spina bifida.

Keywords

Cephalad renal ectopia; subdiaphragmatic kidneys; azygos continuation of IVC; spina bifida; omphalocele.

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Introduction

Renal ectopia is a rare congenital anomaly. Subdiaphragmatic ectopic kidney is one of the rare congenital anomalies, which found to occur once in 22 cases of ectopic kidneys [1]. It is most often discovered incidentally while being investigated for other pathology, as this pathology is frequently asymptomatic. Ultrasonography, excretory urography, computed tomography; with or without contrast enhancement, and magnetic resonance imaging are helpful in differential diagnosis. In most cases, its structure and

functioning are totally normal and thus avoiding surgery or biopsy. Thoracic kidney is considered as the rarest form of renal ectopia than subdiaphragmatic kidneys. There are few case reports of thoracic kidneys have been reported which encountered less than 5% of all renal ectopia [2], however there is no case report of bilateral subdiaphragmatic kidneys has been reported yet, at our best of knowledge. Cephaloid ectopia is usually associated with an omphalocele [3], as in our case. We present a case report of bilateral subdiaphragmatic kidneys, first reported case of its kind in the patient of omphalocele with associated congenital anomalies like azygos continuation of inferior vena cava (IVC), retro-aortic left renal vein and spina bifida. The plain abdominal X-ray, excretory urography, ultrasound and CT urography findings are presented.

Case report

A 5 years old male patient presented with ventral hernia due to omphalocele. Patient was investigated preoperatively for surgical correction of ventral hernia. Ultrasound findings revealed subdiaphragmatic kidney on right side [Fig. 1], however the left kidney was appeared in its normal position. Intravenous urography (IVU) for function of the kidneys and CT urography for association of other abnormalities were advised. IVU revealed normally functioning and excreting bilateral subdiaphragmatic kidneys [Fig. 2].

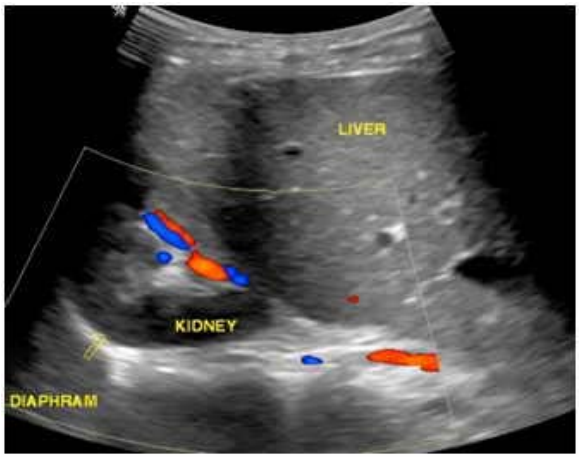

Fig. 1. Ultrasound with color doppler of right upper abdomen shows right kidney between right diaphragm (arrow) and liver suggestive of right subdiaphragmatic kidney.

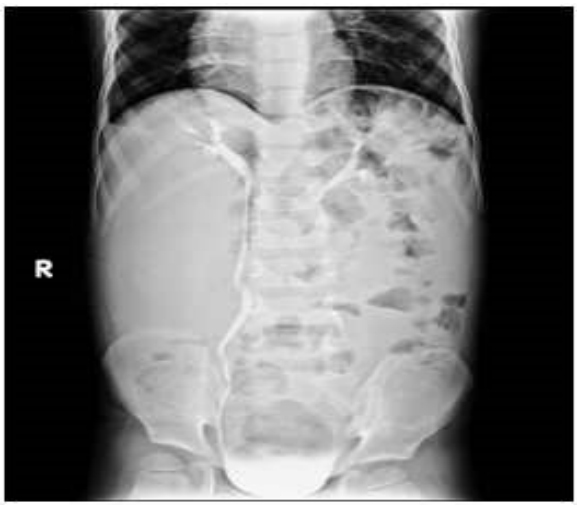

Fig. 2. IVU shows bilateral subdiaphragmatic kidneys.

On CT urography, the subdiaphragmatic position of bilateral kidneys was confirmed [Fig. 3]. There was azygos continuation of IVC with the hepatic veins confluence

directly drained into the right atrium [Fig. 4]. The other anomalies included retro-aortic left renal vein, spina bifida [Fig. 5]. There was mild hepatomegaly with mildly dilated portal vein. The ureters, urinary bladder, bilateral renal arteries and right renal vein were normal.

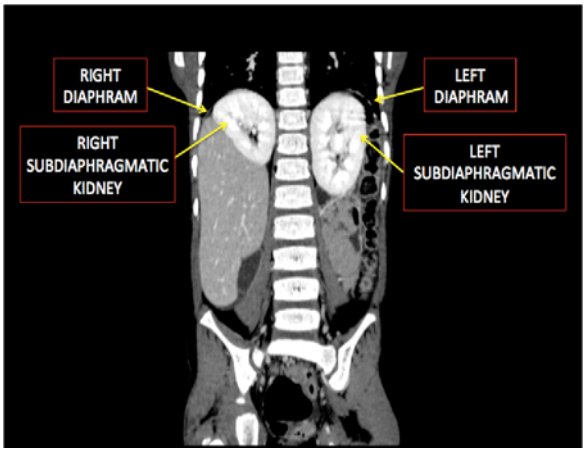

Fig. 3. Contrast enhanced CT scan (coronal reconstructed image) shows bilateral subdiaphragmatic kidneys.

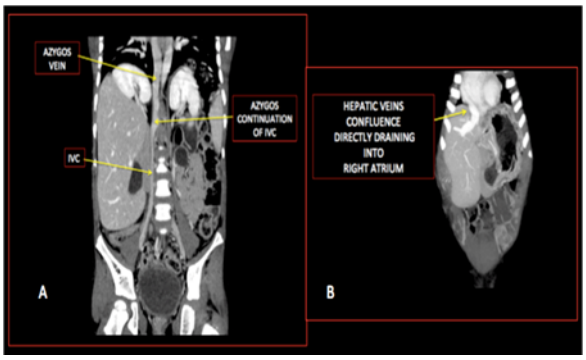

Fig. 4 A and B. Contrast enhanced CT scan (coronal reconstructed images). (A) shows azygos continuation of IVC. (B) shows hepatic veins confluence directly drained into right atrium.

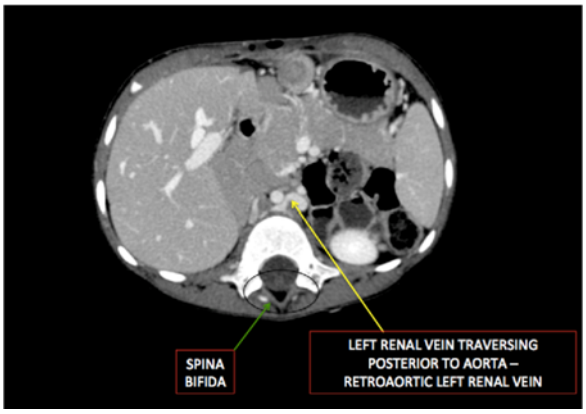

Fig. 5. Contrast enhanced CT scan (axial image) shows spina bifida (black circle). The left renal vein is traversing posterior to aorta, suggestive of retro-aortic left renal vein.

Discussion

Renal ectopia is called when the kidneys are not seen in their normal position of retroperitoneum, against the psoas muscle, on either side of vertebral column. It is thought to occur in approximately 1 in 1,000 births but out of which about 1 in 10 are ever diagnosed. Many of these are discovered incidentally, when a patient is having ultrasonography for other medical condition [4].

The permanent kidneys develop from two sources during development; the ureteric bud and the metanephrogenic cap. The kidneys change their position as development proceeds and gradually ascend up to reach the permanent position opposite the second lumbar vertebra [1].

Subdiaphragmatic ectopic kidney is one of the rare developmental anomalies and, it

occurs 1 in 1000 cases [5]. Subdiaphragmatic type of ectopic kidney is more frequent in males and is more common on the left side [6]. Bilateral subdiaphragmatic ectopic kidneys as seen in our patient are an uncommon and one of the rare developmental anomalies. This anomaly may raise a major problem in differential diagnosis when visualized as pulmonary or mediastinal opacity. The defect is in renal migration while the organogenesis is normal. It is usually associated with omphalocele because the kidneys continue ascent when the abdominal organs herniated in the omphalocele sac [3]. So, the kidneys will reach the subdiaphragmatic region if the migration is prolonged.

It can be either congenital or acquired; the acquired type can be because of injury of the diaphragm in blunt trauma or road traffic accidents [1].

It may be associated with other congenital anomalies of genitourinary system involving absence of vagina, retrocaval ureter, bicornuate uterus, and supernumerary kidney. It may be associated with anomalies of heart, biliary system, musculoskeletal system and bowels [3]. In our case bilateral subdiaphragmatic kidneys were associated with azygos continuation of IVC, retro aortic left renal vein and spina bifida.

In general, bilateral subdiaphragmatic ectopic kidneys are an incidental finding on ultrasound or on chest x-ray. Bilateral subdiaphragmatic kidneys, however rare, must be ruled out in omphalocele patients presented with ventral hernia and should search for other associated anomalies.

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