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Bilateral subdiaphragmatic renal ectopia with associated congenital anomalies: a case report and systematic review of cases

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Abstract

Background: Cephalad renal ectopia is rare. Ectopic kidneys besides being prone to various pathologies are occasionally associated with other congenital abnormalities. To the best our knowledge, at the time of this writing, only ten cases of bilateral subdiaphragmatic renal ectopia had been reported.

Case presentation: We present a rare case of bilateral subdiaphragmatic ectopic kidneys incidentally discovered during evaluation of the abdominal pain. In addition, our patient had associated anomalies of the liver, spleen and mesocardia.

Conclusions: Recognition of this condition is important for accurate diagnosis, surgical and/or intervention planning, as well as identifying other associated anomalies.

Keywords: Ectopic kidney, Cephalad renal ectopia, Mesocardia, Subdiaphragmatic kidney, Omphalocele

Background

Renal ectopia is not uncommon; it is estimated that renal ectopia is seen in 1:900 postmortem examinations and 1:3000 radiological imaging [1]. However, cephalad renal ectopia is rare; for example, intrathoracic or subdiaphragmatic kidneys have estimated incidence of 1 in 22,000 [1]. Importance of imaging is not only to detect renal ectopia, but also to evaluate frequently associated other genitourinary and non-genitourinary congenital abnormalities. There is higher incidence of urinary abnormalities like vesicoureteral reflux or hydronephrosis even in asymptomatic individuals. Ectopic kidneys may have reduced renal function and are often prone to infection, urolithiasis, trauma and malignancies [2–5]. This condition may also pose a challenge during surgery and interventional procedures [6, 7]. Cephalad ectopic kidneys in intrathoracic or subdiaphragmatic locations may mimic mass or simulate pathology on chest radiographs [8–10]. To the best our knowledge, at the time of this writing, there were only ten reported cases of bilateral subdiaphragmatic renal ectopia, but none were associated with mesocardia [11–15]. In this article, we present a case of incidentally discovered bilateral subdiaphragmatic ectopic kidneys with abnormal shape of the liver, and spleen as well as association of mesocardia.

Case presentation

A female in her thirties presented with ongoing episodic epigastric pain after eating and bloating for 10 years. An abdominal ultrasound was first performed which showed cholelithiasis, hepatosplenomegaly and incidental 1.5-cm hypoechoic lesion in the right lobe of liver. Left kidney was poorly visualized on sonography. Magnetic resonance imaging (MRI) of abdomen with contrast was performed to characterize the liver lesion, which revealed that the lesion was slightly hyperintense on T2-weighted

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images, with hyperenhancement on arterial phase and isoenhancing to the rest of the liver parenchyma on portal venous phase. The imaging finding was felt benign, likely focal nodular hyperplasia (Fig. 1a, b). Cholelithiasis was again seen without evidence of cholecystitis. Liver was unusual in appearance with focally enlarged left lobe crossing the midline (Fig. 2a). Spleen was abnormally elongated craniocaudally measuring 19 cm (Fig. 2b). Incidental discovery of unusual cephalad location of bilateral kidneys in subdiaphragmatic location was observed (Fig. 2c). Ectopic kidneys showed normal rotation with renal hila projecting medially. No hydronephrosis or renal calculi were seen. Adrenal gland maintained its relationship with kidneys in anteromedial location. Inferior heart was partially visualized on abdominal MRI with cardiac apex in midline suggestive of mesocardia (Fig. 2d). Atrioventricular relation was maintained. Patient was recommended for echocardiogram for further evaluation of cardiac abnormality. Patient's echocardiogram demonstrated no other cardiac abnormality except for mesocardia.

Discussion

Kidneys develop from the metanephric blastema and ureteral bud starting at fifth week of gestation. The kidneys begin their development in the pelvic region and gradually ascend up to their final positions at the level of T12-L3 vertebra by the ninth week of gestation. This is followed by rotation of the axis resulting in medial position of the renal pelvis. Error in migration of ureteral bud to opposite side and induction of contralateral metanephric blastema results in crossed renal ectopia, while failure of ascent of ureteric bud and metanephric blastema after making contact leads to simple renal ectopia (Fig. 3). Multiple hypotheses for renal ectopia include ureteral bud maldevelopment, defective metanephric blastema, in utero vascular anomalies of kidney at different stages of development and genetic or teratogenic causes. While the etiology of cephalad renal ectopia is not definitive, the condition is thought to be caused by the premature accelerated ascent of the kidneys. If the ascent happens before the diaphragm closes (eight weeks of gestation), the result is intrathoracic kidney. But if the diaphragm closes earlier, then the ascent will result in subdiaphragmatic kidney [1, 16]. Postnatal cases of cephalad renal migration from infra-diaphragmatic location to both subdiaphragmatic and intrathoracic locations have been reported [17, 18]. Most of the reported cases of subdiaphragmatic kidneys had normal renal function [9, 12, 17, 19, 20]. The chances of infection and its complications are less often seen in cephalad renal ectopia compared to caudal renal ectopia due to good drainage in cephalad renal ectopia [21] (Table 1).

Subdiaphragmatic renal ectopia can be isolated or associated with congenital abnormalities. Literature review shows association of omphalocele with cephalad renal ectopia [12, 13]. Presence of omphalocele with herniation of liver and bowel loops probably facilitates cephalad position of the kidney. Also, eventration of diaphragm was observed in at least three cases of subdiaphragmatic renal ectopia, and again the eventration understandably would favor cephalad ascent of the kidneys [8, 22]. Our patient also had abnormal enlarged left lobe of liver, splenomegaly and mesocardia, a combination never seen before. Hepatosplenomegaly was also seen in reported case of bilateral subdiaphragmatic renal ectopia [15]. Most cases of subdiaphragmatic renal ectopia were associated with omphalocele or eventration, conditions that would favor cephalad renal ectopia simply due to availability of space. Our patient also had history of omphalocele repair.

There are evidences to suggest coexistence of cardiac anomalies in patients with renal anomalies and vice versa [26]. Cardiovascular anomalies in cases of

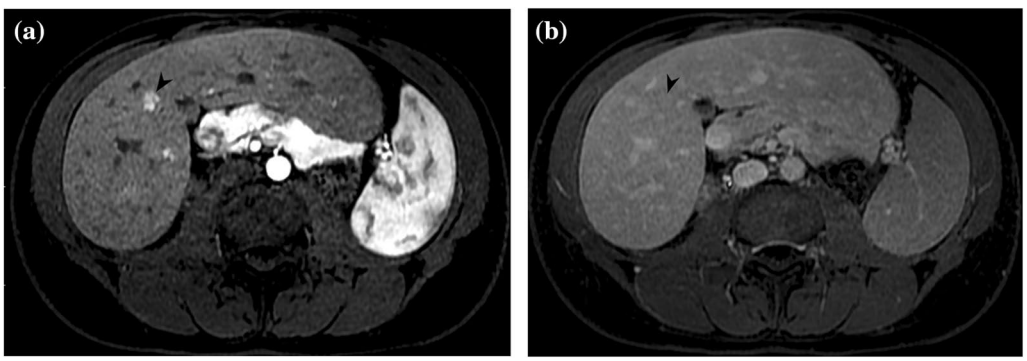

Fig. 1 Axial postgadolinium MRI of abdomen **a** arterial phase demonstrating hyperenhancement, **b** portal venous phase showing lesion (black arrowhead) isoenhancing to rest of the liver parenchyma

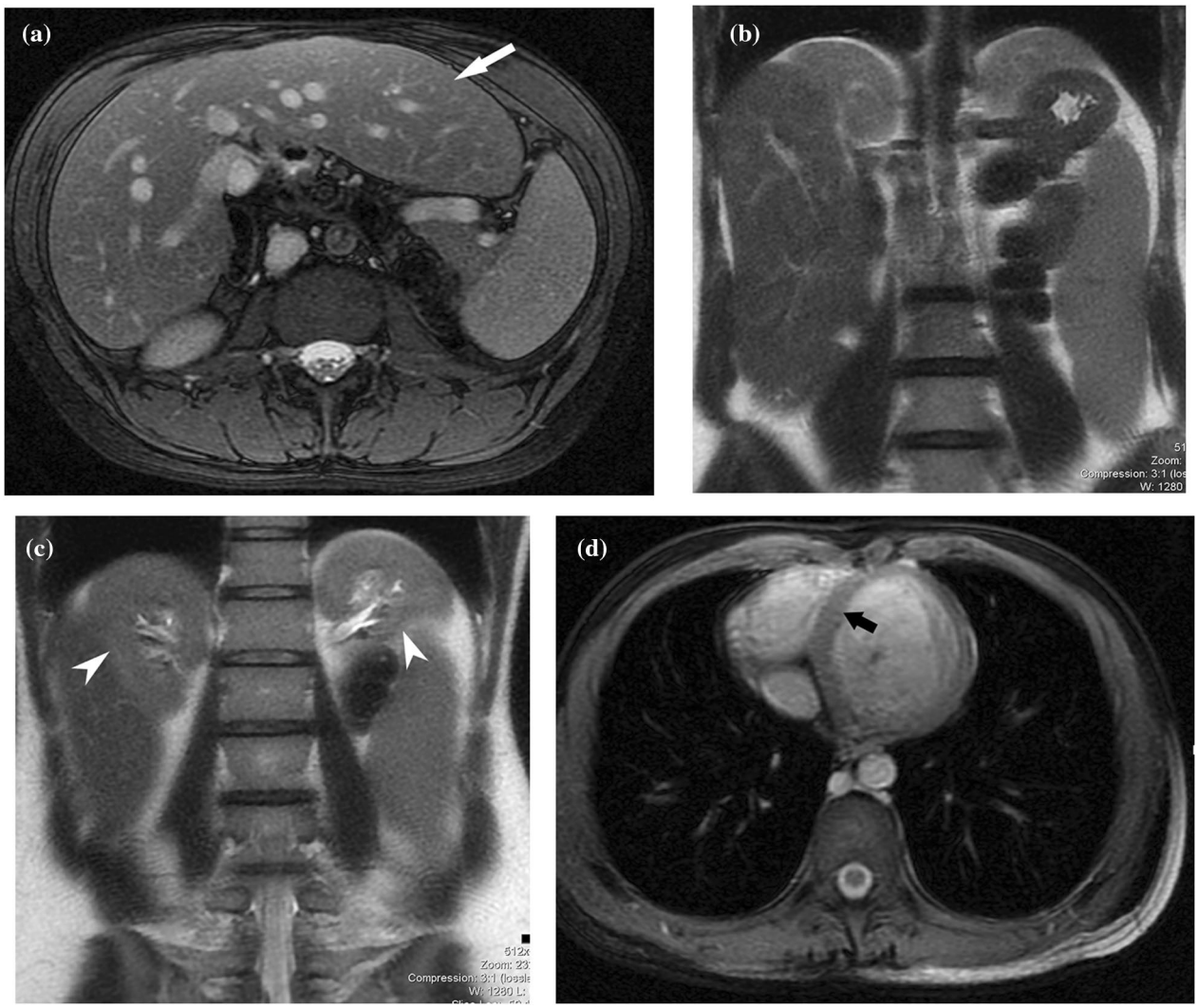

Fig. 2 **a** Axial T2-weighted fat-saturated MRI of abdomen showing abnormally enlarged left lobe of liver crossing midline (white solid arrow), **b** coronal T2-weighted MRI of abdomen showing enlarged spleen, **c** coronal T2-weighted MRI of abdomen showing bilateral subdiaphragmatic kidneys (white arrowheads), **d** axial T2-weighted MRI of abdomen showing interventricular septum (black arrow) and cardiac apex in midline directed anteriorly suggestive of mesocardia

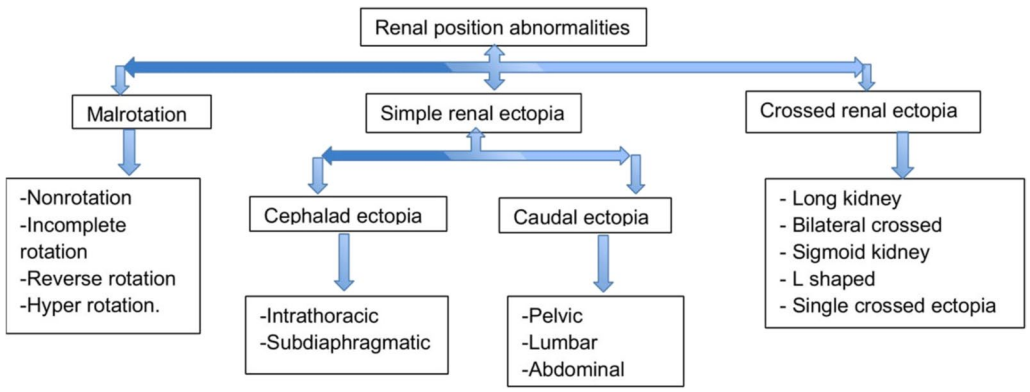

Fig. 3 Different types of renal malposition

Table 1 Systematic review of congenital subdiaphragmatic renal ectopia cases published (based on PubMed and google scholar search for "subdiaphragmatic renal ectopia" on February 16, 2022, as well as case reports in any languages found during the literature review process)

Year	Author	Age	Gender	Presenting symptoms	Laterality	Associated congenital anomaly
1949	Baurys et. al. [19]	NA*	M	NA	Left	NA
1955	James et al. [19]	NA	M	NA	Left	NA
1972	Spillane et al. [22]	4 years	M	Pertussis	Left	Right diaphragmatic eventration
1977	Strakosch et al. [23]	78 years	F	Abdominal pain	Right	unknown
1980	Kaur et al. [19]	55 years	F	Pain hypochondrium	Right	None
1987	Aliotta et al. [13]ǂ	12 mo			Bilateral	Dextrocardia, absent left arm and spleen, endocardial cushion defect, Omphalocele
		18 days			Right	Omphalocele
		3 year			Right	Gastric outlet obstruction, mid-gut malrotation, Omphalocele
		3 weeks			Right	Trisomy 18, Omphalocele
		1 week			Bilateral	Omphalocele
		15 days			Bilateral	Omphalocele, Tetralogy of Fallot
		30 mo			Bilateral	Pulmonary hypoplasia, Omphalocele
1995	Kundu et al. [24]				Left	NA
1999	Attia [8]	38 years	F	Chest pain	Right	Eventration of right hemidiaphragm, bifid ureter and duplication of left renal pelvis, patent ductus arteriosus
1999	Louzir et. al. [9]	72 years	M	Incidental opacity in chest radiograph	Left	None
2004	Muntaha [25]	14 years	F	Abdominal pain	Left	NA
2014	Utanğaç et. al. [20]	22 years	F	abdominal pain	Left	None
2016	Parmar et al. [12]	5 year	M	Ventral hernia secondary to omphalocele	Bilateral	Omphalocele, azygos continuation of inferior venacava, hepatic vein confluence draining directly to right atrium, retro-aortic left renal vein and spina bifida
2016	Zolotas et. al. [17]	3.5 mo	M	Asymptomatic	Right	None
2018	Peng et al. [14]	55 years		Abdominal pain	Bilateral	Primary anterior inferior venacava
2019	Hirayama et al. [10]	76 years	M	Fever	Right	Eventration of diaphragm
2019	Divjak [11]	35 years	F	Intestinal obstruction	Bilateral	unknown
2021	Elshetry [15]	71 years	M	Pain abdomen	Bilateral	Hepatosplenomegaly
2022	Our case	31 years	F	Pain abdomen	Bilateral	Hepatosplenomegaly, mesocardia, omphalocele

*NA- Not available

ǂ- Most of the diagnoses were based on intravenous pyelogram and may not represent true subdiaphragmatic renal ectopia

subdiaphragmatic renal ectopia without omphalocele were patent ductus arteriosus and primary anterior inferior vena cava. In our case, mesocardia was seen, a condition in which baso-apical or longitudinal axis of the heart lies in the midsagittal plane without definite cardiac apex. Atrioventricular relation may be normal or altered in different types of mesocardia. Additionally, these patients may have various abdominothoracic situs as well as other congenital cardiac abnormalities. However, mesocardia with dextro bulboventricular cardiac looping in patients with situs solitus is a normal variant and not associated with other congenital cardiac abnormality, similar to our case. There may be other associated congenital abnormalities of spleen (asplenia, polysplenia and accessory spleen) in patients with mesocardia often determined by situs [27, 28]. In our case, no other abnormality of spleen was present except for splenomegaly.

Clinical presentation in renal ectopia may be non-specific, or patient may remain asymptomatic [29]. Ectopic kidneys particularly intrathoracic and subdiaphragmatic kidneys can pose diagnostic challenges by mimicking mass particularly in chest radiographs [8–10]. Similarly, unusual location of kidneys can be a source of error in nuclear studies if one is not aware of possible cephalad renal ectopia [30, 31]. Having knowledge of this condition from prior imaging may be helpful in decreasing patient's apprehension, preventing inaccurate diagnosis and need for further imaging work-up. Additionally,

surgery and percutaneous intervention may be challenging in patients with these conditions which may have to be tailored based on imaging appearance of ectopic kidney [6, 32].

Conclusions

Bilateral subdiaphragmatic renal ectopia is one of the rarest forms of positional renal anomaly which may be isolated or associated with other congenital anomalies. It is important to be aware of abnormal locations of the kidneys and associated congenital anomalies which may pose not only diagnostic challenges but also help in decision making for surgery or intervention.

Abbreviation

MRI: Magnetic resonance imaging.

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Author contributions

RKC was major contributor for manuscript preparation, literature review and editing. ML prepared manuscript and performed literature review. NP prepared the manuscript, reviewed the literature and helped in editing. EG was major contributor for systematic literature review, manuscript preparation and editing. SS had conception of idea, performed review of the literature and helped in editing. JP had conception of idea and contributed to manuscript review and editing. All authors read and approved the final manuscript.

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Availability of data and material

Not applicable.

Declarations

Ethics approval and consent to participate

Non-identifiable images of the patient have been only used. Careful attention has been provided to make sure that no patient identifiable information is in the images provided.

Consent for publication

Informed consent from the patient has been obtained, and signed consent was provided by the patient.

Competing interests

The authors declare that they have no competing interests.

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