Case Report :

Right thoracic kidney – A rare case report in a female patient.

Saikarthik Jayakumar

Assistant Professor of Anatomy, Department of Maxillofacial Surgery and Diagnostic Sciences, College of DentistryMajmaah University, Al Majmaah 11952

Received on Dec 28, 2023, accepted for publication on March 02, 2024, dx.doi.org/10.5455/mjhs.2024.02.014

Corresponding author:
Saikarthik Jayakumar

SAssistant Professor of Anatomy, Department of Maxillofacial Surgery and Diagnostic Sciences, College of DentistryMajmaah University, Al Majmaah 11952 , Ph: 966 531272304 - Email: s.jaya@mu.edu.sa

OPEN ACCESS Distributed under Creative Commons CC BY-NC-ND 4.0

Abstract

Ectopic kidneys are one the most common forms of renal anomalies. However, intrathoracic kidneys are extremely rare, especially on the right side. Reporting of such cases is relatively very less common in the literature, particularly in the case of females. The current case is an accidental finding noted in a 56-year-old female who came with complaints of diffuse abdominal pain. An initial ultrasound abdomen revealed an absence of the right kidney in the right renal fossa and computer tomography done later revealed a right intrathoracic kidney with a higher origin of the right renal artery (T12 vertebral level). The right ureter was longer and opened into the bladder normally. The author presents this case because of its anatomical, developmental, and clinical importance.

Keywords:
Thoracic kidney, Computed tomography, Angiography, Congenital Diaphragmatic Hernia.

Introduction

The thoracic kidney is one of the rare forms of congenital malformation with a reported incidence noted 1 in every 10,000[1]. Some of the congenital malformations include pelvic kidney, fused kidney (crossed and uncrossed), renal agenesis (unilateral or bilateral), congenital polycystic kidney disease, and ectopic kidneys[2]. Among all the renal ectopia, intrathoracic kidney contributes to less than 5% of the total number of reported cases with left side occurrence being more common[3]. It is also more common among males than females[4]. In the majority of cases, this anomaly is an accidental finding in routine radiography as it is almost asymptomatic[5]. The thoracic kidney may be classified into i) renal ectopia with eventration of diaphragm ii) diaphragmatic hernia (congenital/ acquired) iii) rupture of the diaphragm due to trauma iv) thoracic kidney with closed diaphragm[6].

The present case was an accidental find-

ing that was diagnosed with a right-sided intrathoracic kidney in a female patient. As such an occurrence is quite rare, the present case study is being reported. Ethical approval from the institution and consent from the patient was obtained (MMCRI/36/C19).

Case presentation

A 56-year-old female patient was referred to the radiology department of Madha Medical College Hospital, Chennai, India with the chief complaint of diffuse pain in the epigastric region. There were no other chief complaints. This patient was referred to ultrasonography (USG) abdomen for evaluation.

Investigation:

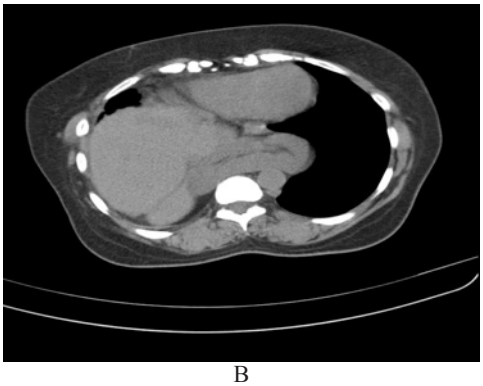
Ultrasound (USG) abdomen screening was performed, and it was noted that there was an absence of the right kidney in the right renal fossa. The left kidney was apparently normal and in situ. No familial history or trauma was reported. After a complete scanning of the abdomen, the right kidney was not traceable. This was informed to the patient and further investigation was advised.

Computed tomography (CT)- Abdomen was performed. Eventration of the right dome of the diaphragm was noticed in the scout image and a herniated thoracic kidney was suspected. A plain CT scan showed a soft tissue mass in the posterior mediastinum (Figure 1A to H).

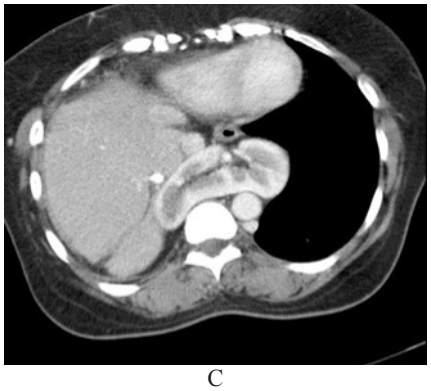
With the patient's renal function test



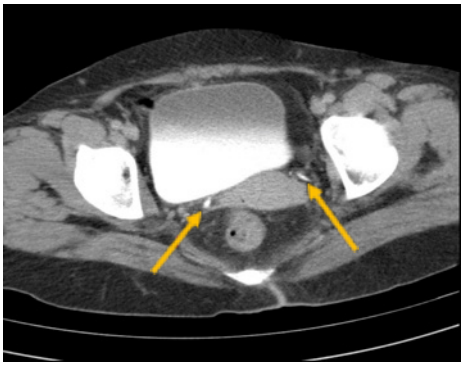
A



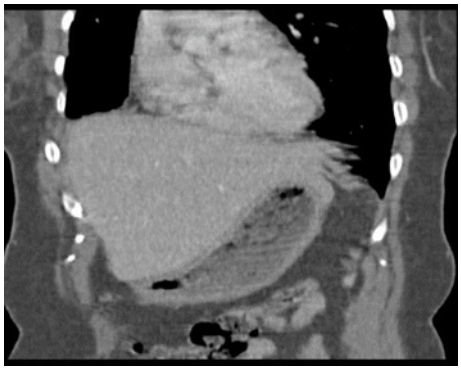
B



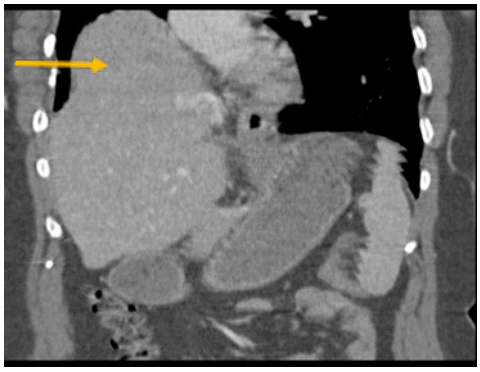
C



D



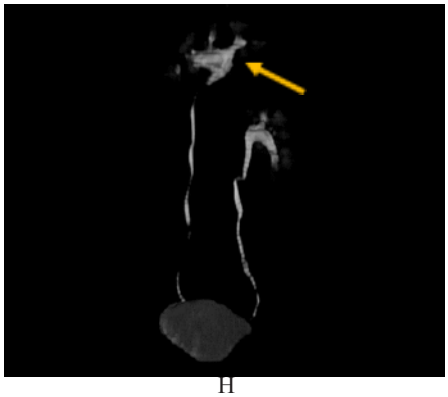
E



F



G



H

Figure 1 A to H show a series of CT scan images of this case. Figure 1A is the scout image. Figure 1B is plain CT at the level of the T8 vertebra. Figure 1C is a contrast image at the level of the T7 vertebra. Figure 1D is a contrast image of the lower pelvis. The orange arrow indicates bilateral ureters. Figure 1 E and F are coronal reformat images. The orange arrow in Figure 1F indicates the herniated portion of the posterior part of the liver into the thorax. Figure 1G is a coronal minimum intensity projection. Notice the severe hypotrophy of the right lung with compensatory enlargement of the left lung. Figure 1H shows the 3D reformat image. Notice the right kidney at the higher level. The position of the hilum of the ectopic right kidney is anteroinferior (orange arrow)

being normal, the contrast-enhanced scan was done to determine the vascularity of this soft tissue. The arterial phase, venous phase, as well as delayed phase, were taken to visualize renal blood vessels, renal tissue enhancement, and ureter and urinary bladder localization.

Cross-sectional contrast images revealed the presence of the right kidney in the posterior mediastinum due to congenital diaphragmatic hernia (CDH) with no hydronephrosis. The hilum of this ectopic kidney was facing Antero-inferiorly with its long axis almost in transverse axis. The right adrenal gland was absent. The right ureter was longer, and it opened on the ipsilateral side of the urinary bladder. Other findings that were noticed in this case included herniation of the posterior segment of the liver, the ostium of the right renal artery at the level of T12 vertebra (along with coeliac trunk), hypotrophy right lung

with compensatory enlarged left lung, mild compression of the azygos vein, inferior vena cava and esophagus between the hilum of the right kidney and the base of the heart.

Discussion

Typically, kidneys begin their development in the pelvis and subsequently migrate to their usual anatomical position in the upper abdomen. The ascent of the kidneys is preceded by the descent of the gonads into the pelvis. Caudal growth during embryonic development seems to facilitate the migration of the kidneys out of the pelvis, placing them in their final retroperitoneal location in the renal fossa. By the 9th gestational week, they attain their adult position. Throughout the ascent, each kidney acquires its blood supply from nearby vessels, initially from the external and internal iliac vessels. By the 8th week of development, direct vascular supply from the aorta is established. Any irregularity in the origin of the renal arteries may hinder the cephalic migration of the kidneys, leading to renal ectopia[7].

Various factors can interfere with this developmental process, including teratogens, genetic factors, abnormalities in the meeting of the ureteric bud with the nephrogenic blastema for normal nephrogenesis, or maternal disease affecting metanephric development. Such disruptions may lead to abnormal migration of the kidney, resulting in renal ectopia[8]. An unusually higher ascent of the metanephros can cause a defect in the diaphragm, resulting in the subsequent presence of an ectopic kidney in the thoracic cavity[9].

The reason behind the cranial migration of kidneys during embryogenesis is still a topic of debate with different schools of thought. Congenital thoracic kidney may be caused by accelerated cranial migration of the embryonic kidney or delayed diaphragm formation. This suggests that aberrant ascent during embryonic development may lead to ectopic thoracic kidney anomaly[10]. Some studies mention the role of the development of the liver and adrenal gland and the persistence of the nephrogenic cord in the development of renal ectopia[11].

Understanding of the genetic factors contributing to kidney anomalies is continuously evolving. Though many genes are involved in renal anomalies, specific genetic abnormality involved in ectopic thoracic kidney is yet to be proven. Studies have shown that CDH is linked with karyotyping abnormalities in 15q26, 8p23.[1], 4p16.3 deletions, tetrasomy 12 associated syndromes such as Pallister-Killian syndrome, Matthew- wood syndrome, and Donnari syndrome[12].

The right ectopic thoracic kidney is a

rare condition where the kidney is located in the thoracic cavity instead of the abdominal cavity. It represents less than 5% of all renal ectopias and is more commonly found on the left side. The ectopia can be associated with other congenital malformations such as CDH and pulmonary sequestration. Diagnosis is usually made through imaging techniques such as chest X-ray and computed tomography. The literature on this condition is limited, and there is a scarcity of information on the impact of ectopia on kidney function[13]. Though it is a rare phenomenon, intrathoracic kidney must be considered for differential diagnosis in cases of intrathoracic masses. In the majority of cases, the ectopic kidney may be asymptomatic and discovered incidentally through imaging tests. In contrast to other renal ectopias, patients with thoracic kidneys have normal renal function and are usually asymptomatic as was the case in the current patient[14]. In terms of the related renal structures, an elongated ureter is a constant finding along with thoracic kidneys which is also found in the current patient[15]. The level of the renal artery may or may not vary with an elongation of the renal artery observed in certain cases. In this patient, the renal artery arises at the level of T12[16].

The ectopic thoracic kidney can present with various symptoms including severe respiratory problems, upper back pain, signs of hemorrhagic shock, and difficulty in breathing. Other symptoms may include chest pain, bilateral leg swelling, and features of malignant hypertension and hypertensive heart disease. Surgical intervention may be necessary in severe cases, but asymptomatic patients do not require treatment as was the situation in the current case. Surgical intervention is advisable in cases where there is kidney blockage or urine backing up to the kidney or in patients with severe respiratory symptoms[17]. A systematic review of the literature on intrathoracic kidneys found that 68% of cases were symptomatic, with 29% experiencing significant complications such as chronic or end-stage kidney disease. Surgical intervention was required in 26% of cases[18]. Complications of intrathoracic kidneys can include pelviureteric junction obstruction (PUJO), renal pelvis stone formation, renal calculi causing obstruction to the superior pole calyces, and malignancy[19]. However, in cases of Bochdalek hernia associated with an intrathoracic ectopic kidney, the functionally normal ectopic kidneys are left in situ during repair of the diaphragmatic defect without complications[20]. This patient was diagnosed with gastric oesophageal reflux and was prescribed GERD medications. Since the patient had normal renal function and no

associated symptoms of thoracic kidney, no intervention was done.

Acknowledgements

The author would like to thank the patient for the support. The author would also like to thank the Department of Radiology, Madha Medical College Hospital, Chennai, India, for their help during the reporting and post-processing of images.

References

1. Peñafiel-Freire DM, Hernández-Martín S, Iceta-Lizarraga A, Urriza-Yeregui L. Ectopia renal intratorácica. Anales de Pediatría. 2020/02/01/ 2020;92(2):113-114. doi:https://doi.org/10.1016/j.anpedi.2019.01.005
2. Gupta R, Gupta A, Ilyas M, Chouhan KS. Adult right-sided thoracic kidney: A very rare form of renal ectopia. Lung India: Official Organ of Indian Chest Society. 2017;34(4):400.
3. Lozano RH, Rodriguez C. Intrathoracic ectopic kidney: report of a case. The Journal of Urology. 1975;114(4):601-602.
4. Lee C-H, Tsai L-M, Lin L-J, Chen P-S. Intrathoracic kidney and liver secondary to congenital diaphragmatic hernia recognized by transthoracic echocardiography. International Journal of Cardiology. 2006;113(3):E73-E75.
5. Aydin H, Sarici S, Alpay F, Gökçay E. Thoracic ectopic kidney in a child: a case report. The Turkish journal of pediatrics. 2000;42(3):253-255.
6. Pfister-Goedeke L, Brunier E. Intrathoracic kidney in childhood with special reference to secondary renal transport in Bochdalek's hernia. Helvetica paediatrica acta. 1979;34(4):345-357.
7. Moore KL, Persaud T, Torchia MG, Persaud T. The developing human: clinically oriented embryology. Saunders. Elsevier, Philadelphia; 2008.
8. Lerman S, McAleer IM, Kaplan GW. Embryology of the genitourinary tract. Clinical pediatric Urology. Martin Dunitz, Boca Raton, FL; 2002.
9. Carrasco A, Castro R. Right diaphragmatic eventration with an intrathoracic kidney: case report and review of the literature. Case Reports in Surgery. 2018;2018
10. Kemper MJ, Müller-Wiefel DE. Renal function in congenital anomalies of the kidney and urinary tract. Current Opinion in Urology. 2001;11(6):571-575.
11. Natarajan A, Agrawal A, Purandare N, Shah S, Rangarajan V. Rare case of thoracic kidney detected by renal scintigraphy. Indian Journal of Nuclear Medicine: IJNM: the Official Journal

of the Society of Nuclear Medicine, India. 2016;31(3):219.

12. Slavotinek AM. The genetics of congenital diaphragmatic hernia. Elsevier; 2005:77-85.

13. Saez RB, Pereira RC, Lopes RS, Emídio FC, Lazenko I. Intrathoracic kidney. European Journal of Case Reports in Internal Medicine. 2023;

14. Drop A, Czekajska-Chehab E, Maciejewski R, Staśkiewicz GJ, Torres K. Thoracic ectopic kidney in adults. A report of 2 cases. Folia Morphologica. 2003-06-09 2003;62(3):313-316.

15. Shimura S, Yoshida K, Kadowaki K, Ishibashi A, Kobayashi K, Fukuda K. Right thoracic kidney with simple renal cyst: report of a case. Hinyokika kiyo Acta Urologica Japonica. 1990;36(11):1321-1324.

16. Yalcinbas YK, Sasmaz H, Canbaz S. Thoracic left kidney: a differential diagnostic dilemma for thoracic surgeons. The Annals of thoracic surgery. 2001;72(1):281-283.

17. Ilgi M, Kutsal C, Albayrak AT, Kirecci SL, Temel U. A rare malformation of urinary system: Right ectopic thoracic kidney. Urology Annals. 2017;9(3):299.

18. Navaratnarajah A, Barry V, Charif R. Bochdalek hernias associated with intrathoracic kidney: a case report and systematic review of outcomes including renal function. Clinical nephrology Case studies. 2020;8:1.

19. Yeow S. Intrathoracic Kidney–A Case Report and Literature Review. Asian Journal of Research and Reports in Urology. 2020;3(3):1-5.

20. Sarac M, Bakal U, Tartar T, Canpolat S, Kara A, Kazez A. Bochdalek hernia and intrathoracic ectopic kidney: Presentation of two case reports and review of the literature. Nigerian journal of clinical practice. 2018;21(5):681-686.