

THORACIC KIDNEY THROUGH RIGHT BOCHDALEK FORAMEN

Andrea Carla de Souza Contencas¹, Lucas Ribeiro dos Santos, MSc², Márcio Luís Duarte, MSc³, Élcio Roberto Duarte, M.D.⁴

¹ Emergency Medicine and Internal Medicine, Santos Faculty of Medical Sciences, Lusiada University, São Paulo, Brazil

² Physiology and Internal Medicine, Santos Faculty of Medical Sciences, Lusiada University, São Paulo, Brazil

³ Radiology, Webimagem Telerradiologia, São Paulo, Brazil

³ Radiology, Prevent Senior, São Paulo, Brazil

* Corresponding author: marcioluisduarte@gmail.com

Abstract

Ectopic thoracic kidney corresponds to less than 5% of all renal ectopia. An asymptomatic 81-year-old woman performed an x-ray which demonstrated a heterogeneous opacity in the right hemithorax. An ultrasonography showed a thoracic right kidney, and a computed tomography demonstrated the right kidney in her right hemithorax through Bochdalek foramen. Physicians must be aware that asymptomatic patients do not need any treatment or invasive procedures due to this malformation, although surgical interventions may be due in severe cases.

Keywords Anatomy; Kidney; X-ray; Ultrasonography; Tomography, X-Ray Computed.

INTRODUCTION

Urinary congenital anomalies are rather common in the general population, especially kidney ectopy; however, the most common presentation is in the pelvic position, followed by renal fusion.¹ Ectopic thoracic kidney usually presents itself on the left side, mainly due to liver positioning, and occurs more often in males.¹

Thoracic kidney ectopy is characterized by the presence of the organ above the diaphragm and inside the posterior mediastinum. It's usually asymptomatic and preserves renal function; the initial diagnosis is often misleading, through a plain

chest x-ray, presenting as an opacity, simulating pleural effusion, or a mediastinal tumor.²

Our aim is to report this rare malformation diagnosed with three imaging tests, x-ray ultrasonography, and computed tomography (CT), in an elderly patient and familiarize health professionals about the avoidance of, often, unnecessary interventions.

CASE REPORT

Here, we present a case of an 81-year-old woman with a history of hypothyroidism and atrial flutter, under anticoagulation. The patient had a left side mastectomy

four years before, due to breast cancer. At consult, she referred a blunt trauma to the right thorax few weeks before, which became painful. The physical examination of the painful region was normal.

An ultrasound of the painful region was performed that identified the kidney above the diaphragm, in the right hemithorax (Figure 1). Moreover, on her chart, she had a plain chest x-ray that demonstrated heterogeneous opacity in the right hemithorax basis (Figure 1). A CT was performed, that displayed the right kidney in her right hemithorax through a right Bochdalek foramen (Figure 2). Routine laboratory analysis showed a preserved renal function.

The patient was discharged with symptomatic simple analgesic, and had remission of tenderness. Follow-up with imaging techniques, as well as estimated glomerular rate filtration evaluation, was the approach of choice.

DISCUSSION

In order for the kidney to surpass the diaphragm, this superior migration should happen in the first 8 weeks of intrauterine life, as the full diaphragm development ends after this period.³

Embryological basis of such malformation remains unknown. An excessive ascent of kidney during migration from the sacral region to the lumbar has been proposed; this process is completed 40 days after conception. It has been hypothesized that a rapid renal ascent, prior to complete closure of the diaphragm could drive to this anomaly.³ Alternatively, a defective/delayed closure of the pleuro-peritoneal membrane could be the leading predisposing factor;^{3,4} nevertheless, co-existence of intra-thoracic kidney and congenital diaphragmatic hernia is exceptional.

Newer mechanistic insights have implicated adrenal and liver development to distort renal position. Still, none of the described mechanisms are mutually exclusive, as they can coexist.⁴

Ectopic kidney is a moderately frequent abnormality (1 in 900 patients).⁵ It may occur in various patterns, with the most usual being pelvic kidney, crossed fused/unfused renal ectopia; thoracic kidney is a rare presentation, and may be located supra, infra, or trans-diaphragmatic.^[6]

The first report of thoracic kidney dates back to 1940, reaching over 200 reports in the modern literature,⁵ and although renal ectopia is a common form of the disease (1: 900 living births), recent reports show that thoracic ectopia accounts for nearly 2% of all renal ectopias.⁶

The left-sided thoracic kidney is a more common abnormality than the right-sided thoracic kidney (62% x 36%); in 2% of the cases, it is bilateral).⁷ This can be justified, because the spleen has a weaker barrier action than

the liver. Associated anomalies are exceptionally rare⁵, however, a case of right intrathoracic kidney with multiple skeleton defects has been published.⁷

Moreover, both environmental and genetic factors are thought to contribute to the etiology of congenital diaphragmatic hernia (CDH) that leads to herniation. Currently, about 30% of the cases have genetic causes identified;⁸ two genes that encodes transcription factors, GATA4 and NR2F2, are the most cited by multiple studies to cause CDH.⁹ Despite many genetic mutations implicated in congenital kidney and urinary tract anomalies, no precise finding have been related to isolated ectopic kidney.¹⁰

If not recognized in the antenatal stage, it may endure silently for several years until discovered incidentally, as it is usually oligo symptomatic, like our case.^{6,7} However, obstruction or vesicoureteral reflux may be present, which could cause chest pain. Moreover, the presence of the long ureter and renal vessels can exit through the Bochdalek foramen, and so, in 0.25%, a Bochdalek hernia may coexist. Other renal complications, such as nephrolithiasis or infection do not have increased incidence, and in most cases, adrenal glands are normally situated.⁵ The ipsilateral ureter is habitually elongated and not ectopic.⁵

There are many valuable tests in the recognition of thoracic ectopic kidney, which are:^{5,6}

- Radiograph: presents soft-tissue opacity in the lower lung zones, but it is unspecific.
- Intravenous urography: demonstrates the position, function, rotational anomaly, and signs of obstruction.
- Ultrasonography: displays the ectopic kidney through the costo-phrenic angles and above the diaphragm.
- Color Doppler study: describes the course of the ectopic kidney vasculature.
- CT: show the position, function, ureteral passage with the aid of multiplan reconstructions.
- CT angiography: describes the path of ectopic kidney vasculature
- Magnetic resonance urography: indicated in cases of impaired kidney function.
- Magnetic resonance renal angiography: evaluates ectopic kidney's vasculature.

Physician must beware of such condition in patients that perform routine x-rays, as the opacity present in the chest radiograph may be misdiagnosed as a soft tissue opacity, loculated pleural effusion, or lung/mediastinum tumor.⁶

Asymptomatic patients do not require any treatment. In cases of reversible injury, intervention is needed to release the obstruction or repair of vesicoureteral reflux; also, repositioning of the kidney may be performed, along with closure of diaphragmatic defect.^{5,6} When the injury is irreversible, and the kidney is nonfunctional, nephrectomy is the elected procedure.⁶

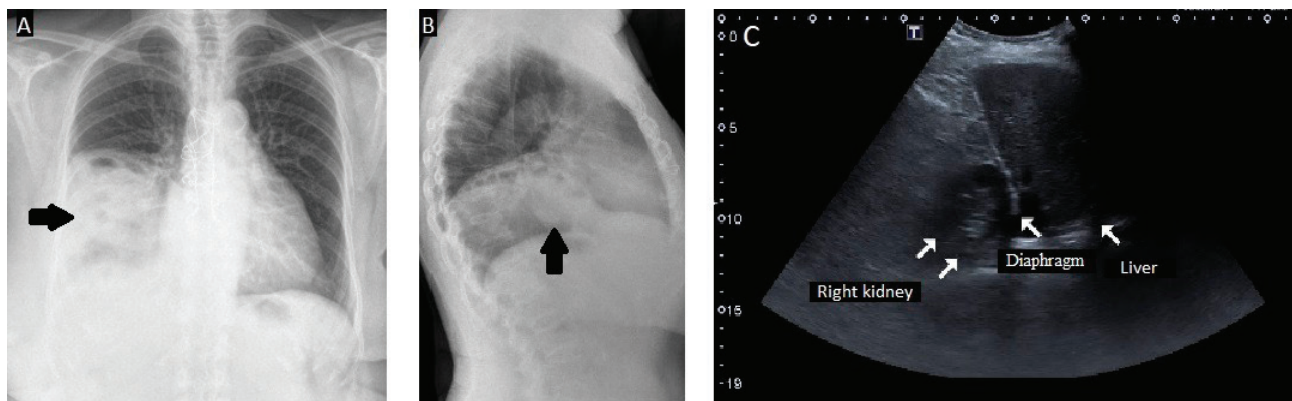


Figure 1

X-ray in AP view in A and profile view in B demonstrating heterogeneous opacity in the right hemithorax (black arrow). In C, ultrasound displaying the right kidney above the diaphragm and the liver.

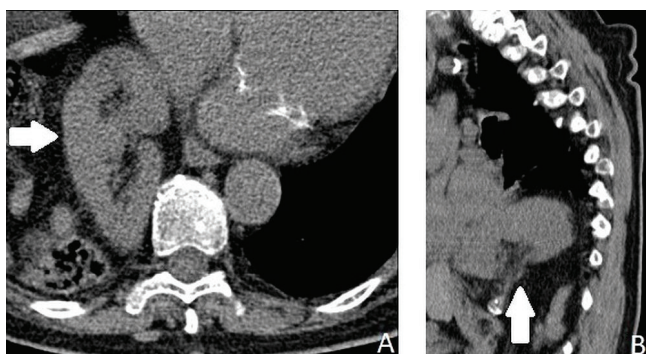


Figure 2

CT scan in axial section in A and sagittal section in B demonstrating the intrathoracic right kidney (white arrow).

CONCLUSION

Thoracic ectopic kidney is an, usually, asymptomatic condition, and the correct diagnosis should prevent patients from unnecessary diagnostic techniques or invasive procedures, as it can be easily diagnosed through imaging techniques such as ultrasound or CT scan, and seldom requires intervention.

Compliance with ethical standards

Funding: No funding was received for this study.

Conflict of interest: The authors declare that they have no conflict of interest.

Ethical approval: All procedures performed in the studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

REFERENCES

1. Demirpolat G, Guney B, Savas R, Alper H. Thoracic Kidney Associated with Partially Intrathoracic Spleen: CT and MRI Findings. *Clinical Radiology Extra.* 2003;58:10–12.
2. Lai CF, Chiang WC, Yang JY, et al. Thoracic kidney and contralateral ureteral duplication--a case report and review of the literature. *Nephrol Dial Transplant.* 2006;21(3):799-801.
3. Magak P, King CH, Ileri E, Kadzo H, Ouma JH, Muchiri EM. High prevalence of ectopic kidney in Coast Province, Kenya. *Trop Med Int Health* 2004; 9: 595-600.
4. Sözübir S, Demir H, Ekingen G, Giivenc BH. Ectopic thoracic kidney in a child with congenital diaphragmatic hernia. *Eur J PediatrSurg* 2005; 15: 206-9.
5. Mensah YB, Forson C. Left thoracic kidney: a rare finding at intravenous urography. *Ghana Med J.* 2010;44(1):39-40.
6. Gupta R, Gupta A, Ilyas M, Chauhan KS. Adult right-sided thoracic kidney: A very rare form of renal ectopia. *Lung India.* 2017;34(4):400-402.
7. Donat SM, Donat PE. Intrathoracic kidney: A case report with a review of the world literature. *J Urol.* 1988;140:131–3.
8. Angulo JC, LopezJI, Vilanova JR, Flores N. Intrathoracic kidney and vertebral fusion: a model of combined misdevelopment. *J Urol* 1992; 147: 1351 – 1353
9. Yu L, Sawle AD, Wynn J, et al. Increased burden of de novo predicted deleterious variants in complex congenital diaphragmatic hernia. *Hum Mol Genet.* 2015;24(16):4764-4773.
10. Kardon G, Ackerman KG, McCulley DJ, et al. Congenital diaphragmatic hernias: from genes to mechanisms to therapies. *Dis Model Mech.* 2017;10(8):955-970.