Adult right-sided thoracic kidney: A very rare form of renal ectopia

Sir,

Congenital anomalies of the urinary tract affect a large sector of human population with renal ectopia being one of the most commonly observed anomaly. Renal ectopia may occur in many forms with most common being pelvic kidney, crossed fused/unfused renal ectopia, and thoracic kidney. Ectopic thoracic kidney forms <5% of all renal ectopia[1] with left-sided thoracic kidney being more common and right-sided thoracic kidney being the rarest form. We are presenting this case of right-sided thoracic kidney because of its rarity and with the aim of familiarizing this situation among clinicians to avoid unnecessary investigations and treatments.

Thoracic ectopia denotes the partial or complete protrusion of kidney above the level of diaphragm into the posterior mediastinum. It is more common in males than females.[2] Usually, it is asymptomatic, and in patients with seasonal cough or mild cold, it may be misdiagnosed as a soft opacity, loculated pleural effusion, or mass lesion on routine chest radiographs. Proper care is to be taken to avoid such mistakes. It is more common on the left side due to less barrier action of the spleen, however, on right side liver acting as a barrier, it is less common, but in our case, it has crossed all the barriers to stay in the right posterior mediastinum.

A 55-year-old male patient presented to chest diseases outpatient department with complaints of mild chest discomfort, cough, and mild fever. He was advised chest radiograph posteroanterior view [Figure 1] which showed a soft opacity in the right lower zone. Sonography of the chest was performed to rule out any pleural effusion which to the surprise revealed that soft-tissue opacity as malrotated and ectopic right kidney in the posterior costophrenic angle with diaphragm separating it from liver [Figure 2] with empty right renal fossa. Contrast-enhanced computed tomography (CECT) of the chest, and abdomen was performed (using Somatom Spirit dual slice computed tomography (CT) scanner by Siemens Healthcare Germany) which revealed a normally functioning ectopic right thoracic malrotated kidney pushing the liver inferiorly. The contrast concentration and excretion were normal [Figure 3]. The left kidney was normal in location with normal uptake and excretion of contrast. No other anomaly was seen. The diagnosis of right-sided thoracic kidney was made and patient informed about the condition.

Renal ectopia describes a kidney that is not located in its usual position thoracic renal ectopia is kidney located in thoracic cavity.
Diaphragm develops by 8 weeks of gestational age of the fetus, so renal ectopia occurs due to abnormally high migration in the first 2 months of intrauterine life. Thoracic kidney is less common on the right side with suggested embryologic basis that right pleuroperitoneal canal (which form the diaphragm) closes earlier than left which makes left-sided hernias and ectopias more common.

Embryologic origin proposes an abnormality in the high migration of the kidney due to delayed mesonephric involution or an abnormality in pleuroperitoneal membrane fusion as causative factors.[3]

Wolfromm was the first to report a case of intrathoracic kidney in 1940. In 1988, Donat and Donat reviewed the world literature and were of the conclusion that intrathoracic kidney was common on the left side (62%) than on the right side (36% as in our case) with only 2% cases having bilateral thoracic kidneys.[4]

Intrathoracic kidney is usually asymptomatic and if not detected during the antenatal period may remain silent for many years until detected incidentally. It becomes symptomatic in less number of cases because although it is ectopic, it functions normally. The symptoms may arise due to obstruction or vesicoureteral reflux and may present as chest pain which may be mistaken for some cardiac pathology. Anatomically, it is usually malrotated with extra-long course of ureter and renal vessels (which exit through the Bochdalek foramen - in our case through right Bochdalek hernia). In 0.25% of cases of thoracic kidney, there may be associated Bochdalek hernia.

The various investigative modalities useful in the confirmation and evaluation of thoracic ectopic kidney include:

a. Chest radiographs: It may show a soft-tissue reniform opacity in the lower lung zones, but it is very difficult to interpret (as happened in our case)
b. Intravenous urogram: It will demonstrate the location, function, rotational abnormality and signs of obstruction (if any)
c. Ultrasonography: It will help in demonstrating the ectopic kidney through the costophrenic angles and above the diaphragm (as in our case)
d. Color Doppler study: It will help in delineating the course of renal artery and renal vein toward the ectopic kidney
e. Contrast-enhanced CT scan: It will perfectly demonstrate the location, position, function, ureteral course with the help of multiplanar reformations (as in our case)
f. CT angiography: It will help in delineation of exact course of renal artery and vein of the ectopic kidney
g. Magnetic resonance (MR) urography: In cases where there are impaired renal function tests, MR urography may be performed for the evaluation
h. MR renal angiography may also play an important role in the evaluation of ectopic kidney’s vasculature.
All these modalities are of great use in the evaluation of thoracic kidney, especially CECT and MR urography.

If asymptomatic, no treatment is required; however, intervention is necessary in symptomatic cases. If reversible damage is there, intervention is required to relieve the obstruction or correct the vesicoureteric reflux. If the damage is irreversible and kidney is scarred, nonfunctional, nephrectomy is the treatment of choice.[5]

**CONCLUSION**

1. Right renal thoracic ectopia is a very rare anomaly (approximately 2% of all renal ectopias)
2. The embryologic basis is due to abnormality in the high migration of kidneys due to delayed mesonephric involution and ectopia occurs within 8 weeks of gestational age with more common on the left side due to delayed closure of pleuroperitoneal canal on the left side than on the right side
3. It is usually asymptomatic but if symptomatic it will present as chest pain caused due to obstruction or vesicoureteric reflux
4. Its symptoms may be confused with that of cardiac or pulmonary origin and may be mistreated, so care is to be taken for proper investigations and treatment
5. The various diagnostic modalities include CT urography, MR urography, color Doppler, CT angiography, MR angiography, and intravenous urogram
6. The treatment procedure depends on the stage of the disease – if reversible, relieve the cause and if irreversible, nephrectomy is the treatment of choice.

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