Unusual association of diseases/symptoms

Incidentally diagnosed right-sided intrathoracic kidney

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Summary

Ectopic kidney is a well-known congenital anomaly. Intrathoracic kidney, as a form of renal ectopia, is an extremely rare entity. A 62-year-old man presented to the department of radiodiagnosis for evaluation of urinary problems. On the basis of ultrasonography, he was diagnosed to be having intrathoracic kidney, which was confirmed by CT scan. We present the details of the patient and brief review of the relevant literature.

BACKGROUND

Ectopic kidney is a well-known congenital anomaly. In most of the patients, the position of the kidney is inferior to the normal position either in the pelvis, iliac or abdomen. Intrathoracic kidney as a form of renal ectopia is an extremely rare entity, which has been reported in about 0.5–5% of cases.¹ It is discovered incidentally in adults on routine chest radiography, presenting as a well-demarcated mass at the base of the lung.²

In humans, three slightly overlapping kidney systems are formed in a cranial to caudal sequence during intra-uterine life—pronephros, mesonephros and metanephros.³ Pronephros is present in the cervical region, mesonephros in the thoracic and upper lumbar and metanephros in the lower lumbar region. Of these, metanephros is the definitive kidney. Kidney, after its formation from the metanephros, ascends up to reach its final position.³ Any aberration in this path may lead to ectopic kidney.

We encountered a patient who was incidentally diagnosed to be having an intrathoracic kidney. Being an extremely uncommon entity, it is being reported with a brief review of the relevant literature.

CASE PRESENTATION

A 62-year-old man presented with complaint of difficulty in micturition for 7 months, along with frequency and hesitancy. There was no other complaint. Digital rectal examination was suggestive of an enlarged prostate and a provisional diagnosis of benign prostatic hyperplasia was made. He was referred for ultrasonography (USG) of the abdomen.

INVESTIGATIONS

On USG abdomen, the size of prostate was 78 g. However, the right renal fossa was empty. On carefully searching for the right kidney, it was seen in the thoracic cavity just above the right dome of diaphragm in the posterior mediastinum. There was no evidence of hydronephrosis. Besides the ectopic right kidney, the rest of the USG abdomen study was normal. There was no evidence of diaphragmatic hernia, and there was no history of trauma.

To know the exact location and relations of the kidney, the patient underwent CT scan of the thorax, which revealed the presence of right kidney on the ipsilateral side in right paravertebral space (figures 1–3).

TREATMENT

As the patient was asymptomatic, no intervention was performed in him.

DISCUSSION

Intrathoracic renal ectopia denotes either a partial or complete protrusion of the kidney above the level of the diaphragm into the posterior mediastinum. In majority of reported cases of renal ectopi, the kidney is not in the pleural space, and the renal vasculature and ureter enter and exit through the foramen of Bochdalek.⁴

The mechanism by which the kidney migrates to a superior position is not known. The physiopathology and embryonic background of the intrathoracic renal ectopia remains unclear. At the end of the eighth week of intra-uterine life, the kidney reaches to its adult location. At this time, the pleuroperitoneal membrane separates the

Figure 1 CT image showing intrathoracic right kidney with the presence of contrast in the pelvi-calycal system. A simple exophytic cyst is also visible.

Figure 2 CT image showing normally placed left kidney. The right ureter is visible as a contrast-filled tube. The right kidney is absent from its normal abdominal place.

pleural cavity from the peritoneal cavity.⁵ Some believed that maldevelopment or delayed closure of pleuroperitoneal membrane permits the kidney to continue its ascent superiorly to appear in the chest. This theory was, however, not considered tenable, as the incidence of thoracic kidney associated with congenital hernia is less than 0.25%. Furthermore, there are only few cases of thoracic kidney associated with an intact diaphragm.^{6,7} Others have suggested that the final location of the kidney is related to the development of the adrenal gland. This mechanism was also not accepted, as the adrenal gland can be located above or below the kidney in superior ectopia.⁷

The most likely acceptable theory is the interaction between mesonephros and metanephros. Normally, the metanephros migrates cranially, and mesonephric tissue involutes in the opposite direction. Delayed involution of mesonephric tissue causes extension of the renal tract, which is considered to be the cause of malposition.¹

Intrathoracic kidney has been classified into four types: (1) thoracic renal ectopia with eventration of the dia-

phragm; (2) thoracic renal ectopia with diaphragmatic hernia (congenital and acquired); (3) traumatic rupture of diaphragm with renal ectopia and (4) thoracic renal ectopia with closed diaphragm. In our patient, the ectopia was of type IV.⁸ The occurrence is more on left side (66%) as compared with the right side.⁴

The diagnosis is mostly incidental, which was also seen in our patient. On chest x-ray, it appears as a well-demarcated mass at the base of the lung.⁹ USG may demonstrate the presence of ectopic kidney in the thoracic cavity, where it may be recognised by its typical appearance. Intravenous urography could be used to confirm the diagnosis; however, CT scan is now considered to be the mainstay of diagnosis. The CT urogram can diagnose function, associated anomalies, stones and visualise vascular and ureteral anatomy. Recently, renal cortical scintigraphy has been recommended to confirm function and to exclude any associated renal anomalies.¹⁰ MRI has also been used to diagnose it.¹¹

Thoracic kidneys are usually asymptomatic and function normally. The stretched ureter provides good drainage, and infection and other complications are uncommon. No specific treatment is necessary as renal or pulmonary symptoms are exceedingly rare and only documentation for future reference is necessary.⁴

Learning points

- ▶ Intrathoracic kidney is a rare condition.
- ▶ It needs to be considered as a differential diagnosis for a posterior mediastinal mass.
- ▶ Radiological diagnosis is confirmatory.
- ▶ The prognosis of the condition is good.

Competing interests None.

Patient consent Obtained.

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Figure 3 CT reconstructed image showing intrathoracic right kidney with contrast-filled normal pelvi-calyceal system.

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