CASE REPORT

Ectopic intrathoracic kidney: A case report and literature review

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Available online 30 April 2013

Summary Intrathoracic kidney is a rare congenital abnormality with the lowest frequency among all renal ectopias. We report the case of a 20-year-old asymptomatic female patient who came to our institution for an evaluation of an incidentally noted right lung base opacity on a plain chest radiograph. The subsequent computed tomographic scanning led to the diagnosis of thoracic renal ectopia. In this article, we discuss the relevant clinicoradiological findings along with a review of the literature.

Introduction

Renal ectopia refers to a kidney situated in any location other than the renal fossa. Ectopic kidneys are thought to occur in approximately 1 in 1000 births, but only about 1 in 10 of these is ever diagnosed. With a prevalence rate of less than 0.01%, intrathoracic kidneys represent less than 5% of all renal ectopias; indeed, it has the lowest frequency rate among all renal ectopias.1–3 It has therefore a reported incidence of less than 5 per 1 million births.4 It is generally seen as an incidental finding detected on chest radiograph simulating a posterior mediastinal mass and mandating further evaluation.5 We present a similar case encountered in our routine clinical practice, emphasizing the importance of cross-sectional imaging in diagnosing this relatively benign condition.

Case report

A 20-year-old female patient was referred to our institution because of an incidentally detected chest lesion. Chest
radiograph revealed an ill-defined radio-opacity in the lower part of the right hemithorax (Fig. 1). There was neither a history of any trauma or operative procedure nor any complaint of previous respiratory or urological disease. Results of her physical examination were unremarkable. All the relevant blood and urine tests yielded normal results. The computed tomography (CT) scan elegantly demonstrated the presence of ectopic reniform structure in the intrathoracic location on the right side (Fig. 2). Post contrast images showed normal contrast excretion and nondilated pelvicalyceal system, indicating normal functioning of the intrathoracic kidney (Fig. 2). The patient was discharged and followed-up on an outpatient basis.

Discussion

Intrathoracic kidney is a partial or complete protrusion of the kidney above the hemidiaphragm into the posterior mediastinal compartment of the thorax. The first case of thoracic kidney was diagnosed by Wolfromm in 1940 using retrograde pyelography. Since then, very few such cases (≈ 94) have been reported. This condition shows male predominance and occurs more commonly on the left than on the right side. Ten percent of cases are bilateral. It is noteworthy that in all cases, the kidney is located in the thoracic cavity and not in the pleural space, with renal vessels and ureter typically exiting the thorax through the foramen of Bochdalek.

Various mechanisms have been thought to be responsible for intrathoracic kidneys such as accelerated ascent
of the kidney, delayed closure or maldevelopment of the pleuroparietal membrane, effect of the developing liver and adrenal glands, and the persistence of the nephrogenic cord.\textsuperscript{12,13} During embryogenesis, the kidneys are initially situated in the pelvis; then, they ascend into the abdomen as the caudal portion of the embryo grows relative to cranial. Ascent stops when the kidneys reach the adrenals. In actuality, both kidneys are physically hindered from higher ascension predominantly by superiorly located adrenals and, to some extent, by the liver. Thus, under conditions affecting the development of adrenal glands and liver, the ascending developing kidney may rarely “overshoot” and ascend to a higher location than normal, resulting in thoracic ectopia.\textsuperscript{14,15} However, none of these postulated mechanisms can solely explain all the reported cases.

Most patients with intrathoracic kidneys are asymptomatic and have a benign clinical course. However, anatomically, rotational anomalies (such as hilum facing posteriorly, long ureter, high origin of renal vessels) and medial deviation of lower pole of kidney may be seen. Associated anomalies in other organ systems are extremely rare.\textsuperscript{16,17}

Several methods have been used to diagnose intrathoracic kidney. Plain radiographs are often indeterminate and may confuse this condition with other posterior mediastinal lesions such as Bochdalek hernia, pulmonary sequestration, or neurogenic masses. In the past, intravenous urography was the modality of choice for confirming the diagnosis, but it has been superseded by ultrasonography and CT scan in recent times.\textsuperscript{16,18}

Nuclear imaging also plays an important role in its diagnosis. Tc-99m DMSA (dimercaptosuccinic acid) and Tc-99m DTPA (diethylenetriamine pentaacetic acid) scintigraphy can be used to differentiate an ectopic thoracic kidney from other tissues.\textsuperscript{19} Renal scintigraphy must be performed even if CT and intravenous pyelogram results are normal, because it depicts the kidney function more accurately.\textsuperscript{12}

Treatment is not required in the majority of cases of intrathoracic renal ectopia, except in those associated with other anomalies such as vesicoureteric reflux and obstruction.\textsuperscript{13,18,20}

In conclusion, intrathoracic renal ectopia is a rare clinical entity and is a diagnostic challenge for both clinicians and radiologists. Awareness of this abnormality along with a high index of suspicion may obviate the need for unnecessary investigations and operative procedures.

References

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