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A right thoracic kidney with a pulmonary vascular malformation in an adult

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Abstract

Ectopic thoracic kidneys are rare anomalies, accounting for less than 5% of all renal ectopia. The anomaly is usually found incidentally on a chest radiograph, with no respiratory or systemic symptoms. It usually presents in the left posteroinferior thorax, associated with a congenital diaphragmatic defect or hernia and is more frequent in males than females. Associated anomalies of other organs are rare and inconsistent. We report a case of a 55-year-old female who was referred to our hospital for evaluation of a mass-like lesion in the right chest, identified incidentally on a chest radiograph. Contrast-enhanced chest computed tomography revealed a right ectopic thoracic kidney with a closed diaphragm and an accompanying pulmonary vascular malformation.

Introduction

Ectopic thoracic kidneys are rare, accounting for less than 5% of all renal ectopia [1, 2]. It usually presents in the left posteroinferior thorax associated with a congenital diaphragmatic defect or hernia and most are found in males. Associated anomalies of other organs are rare and inconsistent. We report a case of a right ectopic thoracic kidney with a closed diaphragm and an accompanying pulmonary vascular malformation identified incidentally.

Case Report

A 55-year-old female with a history of pulmonary tuberculosis at 17 years of age was referred to our hospital for evaluation of a mass-like lesion in the right chest, identified incidentally on a chest radiograph. The patient was a current smoker with 2 pack-years and was well with no respiratory or systemic symptoms. She had no history of trauma. The physical examination was unremarkable.

The posteroanterior chest radiograph showed a well-marginated round lesion in the lower right lung, measuring approximately 9 cm along its long axis and 5 cm along its

short axis (Fig. 1A). On the lateral chest radiograph, the lesion was above the diaphragm, located posteroinferiorly in the thorax (Fig. 1B). Chest computed tomography (CT) revealed an ectopic kidney above the right diaphragm in the posteromedial right thorax (Fig. 2A-F) and an accompanying right ectopic adrenal gland (Fig. 2A). There was no evidence of a diaphragmatic defect or hernia (Fig. 2C-F). There was also a highly enhancing vascular structure in the right lower lobe (Fig. 2A-C, white arrow), suggesting a pulmonary arteriovenous malformation. CT pulmonary angiography was planned to evaluate this further, but the patient refused and is being followed regularly as an outpatient.

Discussion

An ectopic thoracic kidney is a rare anomaly, accounting for less than 5% of all renal ectopia [1, 2]. Since the first thoracic kidney was reported in a 43-year-old female patient using retrograde pyelography in 1940 [3], few cases have been reported and its overall prevalence is less than 1/10,000. The anomaly is usually found incidentally on a chest radiograph. It is more common in the left

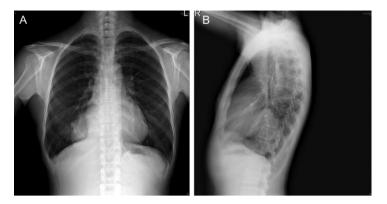


Figure 1. The posteroanterior chest radiograph shows a round, well-marginated lesion with long- and short-axis diameters of 9 and 5 cm, respectively, in the right lower thorax (A). In the lateral chest radiograph, the lesion is above the diaphragm in the posteroinferior area (B).

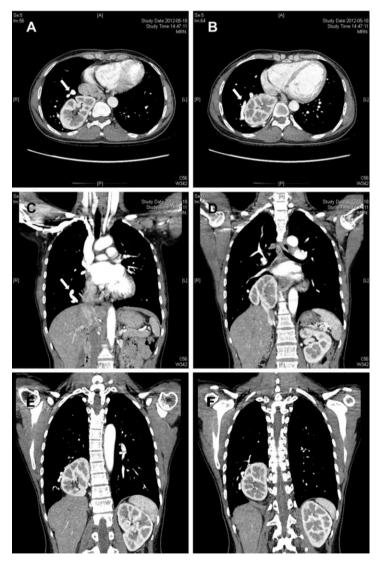


Figure 2. Axial chest CT shows an ectopic kidney and adrenal gland in the posteromedial right thorax with a likely vascular malformation in the right lower lobe (arrowed) (A,B). Coronal chest CT further demonstrates the likely arteriovenous malformation and demonstrates the right hemidiaphragm to be intact (C-F).

posteroinferior thorax, associated with a congenital diaphragmatic defect or hernia and is more frequent in males than females. The ectopic thoracic kidney usually functions normally, and most patients with an ectopic thoracic kidney have no genitourinary symptoms and it only rarely causes respiratory symptoms. Regarding the phenotype, Pfister-Goedeke and Brunier [4] classified the anomaly into four categories: (1) thoracic renal ectopia with a closed diaphragm; (2) eventration of the diaphragm; (3) a diaphragmatic hernia (congenital diaphragmatic defects or an acquired hernia, such as a Bochdalek hernia); and (4) traumatic rupture of the diaphragm with renal ectopia. In our case, unlike other reported cases, the ectopic thoracic kidney was identified incidentally in the right thorax in an adult female with a closed diaphragm on chest CT.

Concurrent anomalies in other organs with an ectopic thoracic kidney are rare and inconsistent; few cases have been reported, including accompanying genitourinary anomalies [5], a wandering spleen, dextrocardia, and patent ductus arteriosus. Our patient had an ectopic thoracic kidney with an ectopic adrenal gland and accompanying abnormal pulmonary vasculature suggestive of a pulmonary arteriovenous malformation, which has not been reported previously.

Various embryologic etiologies of ectopic thoracic kidneys have been suggested. However, it is not clear whether it is due to an abnormality in pleuroperitoneal membrane fusion or in the migration of the kidney due to delayed mesonephric involution. The diagnosis of an ectopic thoracic kidney is easily made using contrast-enhanced CT, intravenous pyelography, magnetic resonance imaging (MRI), or ultrasonography; these can help to differentiate the anomaly from

neoplasms of the thorax or infectious conditions. Treatment for the ectopic thoracic kidney is necessary only if signs of urinary obstruction or associated symptoms are present, and the overall prognosis is good.

In conclusion, our report reminds readers that ectopic thoracic kidneys may be right or left-sided, may be associated with an intact diaphragm and may also be associated with pulmonary arteriovenous malformations.

Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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