Ectopic intrathoracic kidney in a pediatric patient

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Abstract
Renal ectopia refers to a kidney situated in any location other than the renal fossa. It occurs in about 1 in 1000 births, but only 1 in 10 of them have been diagnosed. In this study, we report the case of a female patient, 2 years old, in whom a mass in the left hemithorax was evidenced on the chest radiograph. Ultrasonography showed that the patient’s left kidney was in the chest, the left of the heart, an examination confirmed later by computed tomography. Due to the difficulty in elucidating whether renal ectopy would be secondary to diaphragmatic failure or only to eventration, a nuclear magnetic resonance imaging of the thorax was requested, which proved that renal ectopy was secondary to eventration of the diaphragm. As in the case reported, diaphragmatic eventration is not usually symptomatic, nor does it generate clinical repercussion, and does not require surgical treatment most of the time. Thus, it was decided to keep follow-up at outpatient level. In addition, although rare, intrathoracic kidney should not be overlooked in the differential diagnosis of intrathoracic mass.

Keywords:
kidney, thoracic cavity, congenital abnormalities, infant.

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INTRODUCTION

Ectopic kidney refers to a kidney located at any site other than the renal fossa. It occurs in approximately 1 in 1,000 births, but only 1 in 10 of them were diagnosed1.

The ectopic kidney can be pelvic, iliac, abdominal, intrathoracic, contralateral or crossover2. Of these subtypes, the pelvic is the most frequently reported, and the intrathoracic kidneys are the least, representing less than 5% of the cases, with a reported incidence of less than 5 cases per 1 million births1,3. In the medical literature, up to 2012, only 13 cases of intrathoracic kidney in the pediatric age group had been reported4.

In most cases, intrathoracic kidneys are incidentally discovered as a mass on a chest radiograph5. They are usually asymptomatic, in other cases they can cause respiratory or urinary problems. They can function normally, although they are not in their usual position6.

The intrathoracic kidney is more frequent in males, with a ratio of 2:1. It is usually seen on the left side (61%), but can also be located on the right side (36%) or bilaterally (2%). In all cases, the kidney is located in the thoracic cavity and not in the pleural space, with renal vessels and ureter usually leaving the thorax through the Bochdalek foramen (posterolateral diaphragmatic foramen)1,3.

The normal kidney originates oppositely to the twenty-eighth somite, at the level of the fourth lumbar vertebra, where the ureter joins caudally. In the fetus at term, the kidney has already ascended to the level of the first lumbar vertebra or even to the twelfth thoracic vertebra. This kidney ascension is caused not only by its regular cephalic migration, but also by differential growth in the caudal region of the body. During the initial ascending period (seventh to ninth week), the kidney slides above the bifurcation of the aorta artery and rotates 90 degrees. Its convex edge is directed laterally, not dorsally. The ascension proceeds more slowly until the kidney reaches its final position7.

The diaphragm is completely formed by the ninth week, and the continuous ascension of the kidney produces its intrathoracic umbilication in the presence of the incompletely formed diaphragm. The low incidence of right renal ectopy is explained by the early fusion of the pleuroperitoneal canal on this side and the liver as a barrier8.

Several factors have been suggested as causes of intrathoracic ectopic kidney, such as rapid kidney ascension, late closure or malformation of the pleuroperitoneal membrane, defects in the development of the liver or adrenal glands, and persistence of the nephrogenic cord1.

During the intrauterine period, the following factors may play a role in this etiopathogenesis: folic acid or vitamin A deficiency, exposure to teratogenic drugs, chemical substances, ionizing rays, infections or infestations such as Schistosoma haematobium or malaria3.

The diagnosis of intrathoracic kidney ectopy is still challenging. Formerly, excretory urography was the imaging exam of choice for the diagnosis of this condition, but it has now been superseded by computed tomography and ultrasonography. After diagnosis of ectopia, renal scintigraphy should be performed to assess renal function with greater accuracy1.

Although the initial literature has recommended aggressive treatment, most cases of intrathoracic kidney do not require a surgical approach, leading to increasingly conservative behaviors. The surgical treatment in pediatrics is reserved for cases where there is an association with intestinal hernia or respiratory compromise, especially in children with bilateral intrathoracic kidneys, because there is concern about the growth and development of these kidneys, besides the potential vascular or ureteral obstruction4.

CASE REPORT

I.F.S., 2 years, female, coming from Vicência-PE, came to the emergency department with a history of fever for 4 days, increased abdominal volume for 3 days and vomiting for 2 days. At the initial physical examination, the patient was in good general condition, pale (+/- 4+), bulbous abdomen, painless, with a palpable liver at 4 cm from the right costal border and spleen at 2 cm from the left costal border. We then started to investigate her febrile hepatosplenomegaly.

During the clinical investigation, the chest X-ray revealed an image suggestive of diaphragmatic eventration, and the ultrasound of the abdomen and thorax showed that the patient’s left kidney was in the chest, to the left of the heart. CT scan of the chest and abdomen showed liver and spleen with discretely enlarged dimensions, presence of left intrathoracic kidney in intimate contact with the atrium and left ventricle. The kidney had regular shape, volume, contour and attenuation. As there was difficulty in elucidating whether kidney ectopy would be secondary to diaphragmatic failure or only eventration; we then ordered an MRI of the thorax (Figure 1) for better analysis. This clarified eventration/lobulation of the posterior portion of the left diaphragmatic fault, determining compressive partial atelectasis of the adjacent pulmonary base. A left-side ectopic kidney, in the left thoracoabdominal transition. The left kidney and spleen angle of the colon projected to the base of the left hemithorax.

After diagnosis of kidney ectopy and its proximity to the heart, a transthoracic echocardiogram showed no anatomical and functional alterations of the heart.

The patient was then seen by pediatric nephrology and surgery specialists who, due to the good general condition of the patient and the fact that the renal ectopy findings were not determining any immediate clinical repercussions, opted to maintain follow-up in an outpatient basis, scheduling a renal scintigraphy, in order to evaluate the ectopic kidney function.

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Figure 1. MRI showing an ectopic kidney in the left hemithorax.

DISCUSSION

The intrathoracic kidney is the rarest renal ectopia, with only 200 cases reported in the literature, 13 of them in children. Most of the cases are discovered in the presence of a mass in the thoracic region, as was the case of our patient. As in the case of our patient, there is a discrete predominance of the left side (61%) in relation to the right (36%), being this disorder rarely bilateral. However, there is a great predominance of males in relation to females, which is in agreement with our findings.

Intrathoracic renal ectopia can be classified as: (I) true ectopia; (II) diaphragmatic eventration; (III) diaphragmatic herniation; and (IV) traumatic lesions of the diaphragm. These subtypes have therapeutic and prognostic repercussions. In the case of the patient in question, the MRI showed signs of diaphragmatic eventration, with left kidney and spleen angle of the projected colon for the left hemithorax.

The intrathoracic kidney ratio to the diaphragm may vary from case to case. Ours was accompanied by diaphragmatic eventration, which is caused by insufficient muscle fibers due to paralysis, aphasia or atrophy in the prenatal period and it is a very rare condition. As in our case, diaphragmatic eventration is not usually symptomatic. For this reason, most of the time, it does not require surgical treatment, and therefore, we decided to maintain follow-up of the patient in an outpatient basis.

According to the literature, kidney ectopia is related to multiple systemic abnormalities. In females, genital abnormalities such as agenesis of the uterus and vagina (e.g., Müllerian agenesis and Mayer-Rokitansky-Küster-Hauser syndrome), or unicorn uterus have been associated with renal ectopy. However, during the patient’s hospitalization, no other congenital malformations were evident.

Thus, in the differential diagnosis of intrathoracic mass, even though it is a rare entity, one should think of an intrathoracic kidney, which is usually asymptomatic and can be identified by ultrasonography or chest tomography, as in the case in question.

FINAL REMARKS

Most cases of intrathoracic ectopic kidney appear as a tumor found on chest radiographs requested for any reason other than the suspicion of this anomaly, and does not require specific treatment.

REFERENCES