A three-year old girl with Down's syndrome and an abnormal finding in the chest

Editor: Husn Frayha
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A 3-year-old girl known to have Down's syndrome (trisomy 21) had initially presented at the age of 18 months with a fever of about one month duration. She was found to have hepatosplenomegaly. The complete blood count revealed WBC 91×10^9/L, hemoglobin 5.4 g/dL, and platelets 21×10^9/L. A peripheral blood smear showed 80% blast cells. She was diagnosed as acute myeloid leukemia, French-American-British (FAB) classification M2. She was treated with chemotherapy and responded and came off therapy. She had normal renal and hepatic biochemical profiles during and after treatment.

During an episode of febrile neutropenia due to chemotherapy, her evaluation for the cause of fever included a chest x-ray (Figure 1) and CT (Figure 2), which showed an abnormal finding in the chest.

- What is your differential diagnosis?
- What is the diagnosis?

(Answer on page 354)

Figure 1. Chest x-ray (frontal and lateral views) of the patient.

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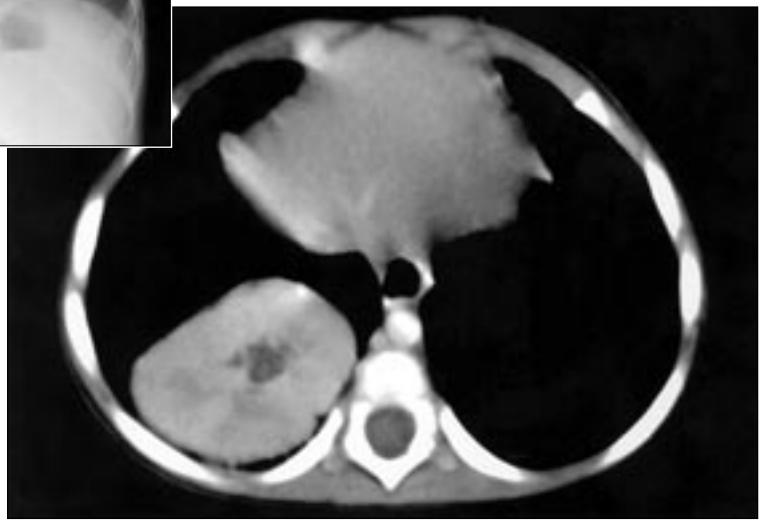

Figure 2. CT scan of the chest.

Diagnosis: Right Intrathoracic Renal Ectopia

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An ectopic kidney is one found in an abnormal position, usually in the abdomen or pelvis in the form of crossed ectopia, crossed fused ectopia or pelvic kidney.[1,2,3,4] Ectopic kidney in the chest is rare and needs to be differentiated from any suspected pathology. In this case, the thoracic kidney was missed on the chest x-ray, but retrospectively, a subtle, triangular opacity projected over the right hemidiaphragm (Figure 1). Ultrasound showed only the lower part of the kidney, behind the liver. Because ultrasound cannot see through the air of the lungs, the interpretation was a small rotated right kidney. CT showed the right kidney surrounded by the right lower lobe parenchyma (arrow, Figure 2). The kidney showed normal shape and perfusion. CT was the only modality that clearly depicted the kidney inside the right hemithorax.

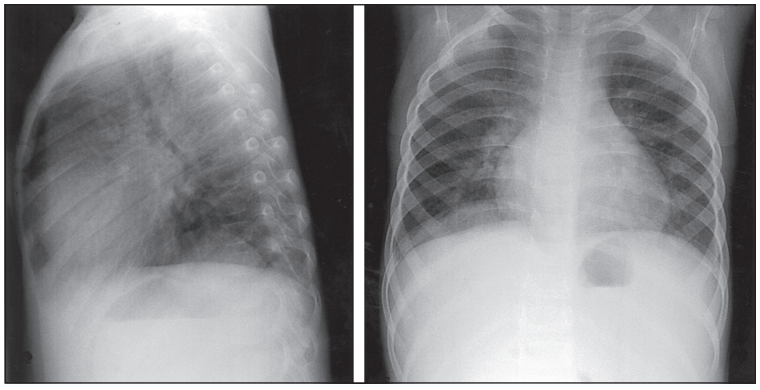

Figure 1. Thoracic kidney as subtle, triangular opacity project over the right hemidiaphragm on chest x-ray.

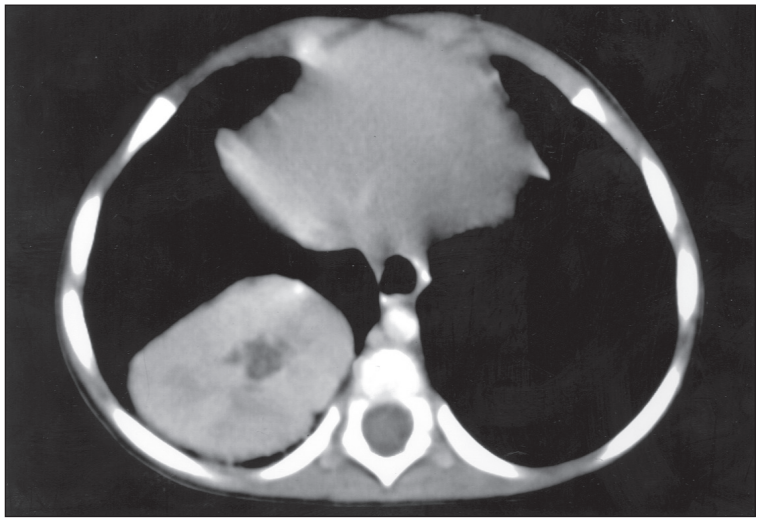

Figure 2. The kidney inside the right hemithorax.

Discussion

The kidney develops in the pelvis in the fifth week of intrauterine life and then ascends, reaching a normal position by the eighth week.[5] If the kidney reaches the thorax, it interferes with development of the diaphragm, resulting in diaphragmatic hernia or eventration of the diaphragm. Thoracic ectopic kidney can be classified by the presence of either (a) a normal diaphragm, (b) eventration of the diaphragm, (c) congenital hernia (Bochdalek), or (d) post-traumatic rupture of the diaphragm. Thoracic renal ectopia is more common in males than females (ratio of 1.7:1), and the majority are on the left side (66%), with only 2% located bilaterally.[1,3] The criteria for diagnosis of ectopic thoracic kidney are rotation anomaly, long ureter, anomalous high derivation of the renal vessel, and medial deviation of the lower pole of the kidney. Ectopic thoracic kidney can be an isolated finding or it can be associated with other congenital anomalies,[3] trisomy 21[1] or an anomaly in the opposite kidney,[6] pulmonary or cardiovascular abnormalities,[3] or skeletal abnormalities.[4]

Plain chest x-ray may show a mass in the thorax, but cannot tell the nature of the mass. On the other hand, ultrasound can show that an ectopic kidney is abnormal, but cannot depict the whole kidney due to the limitation of ultrasound in the presence of air in the lung surrounding the kidney. A renal nuclear scan can recognize the intrathoracic mass as the kidney and provide information on its function. However, the modality of choice is the CT scan, which can show the nature of the mass, the perfusion and the excretion of the intravenous contrast given during the study. The rare occurrence of intrathoracic ectopic kidney should not make us forget to include it in the differential diagnosis of the intrathoracic mass.

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