Intrathoracic kidney, dextroposition of the heart, right upper and middle lobe hypoplasia of lung and pulmonary hypertension

***Mohammed Abdul Wasiq**[1], **J B K Prusty**[1], **Anuspandana Mahapatra**[1], **Mrutunjay Dash**[1]

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(Intrathoracic kidney, dextroposition of heart, pulmonary hypoplasia, mesonephros, metanephros)

Introduction

Intrathoracic kidney was first described by Campbell in 1930[1]. Intrathoracic kidneys constitutes less than 5% of all renal ectopias and have the lowest frequency rate in comparison to other forms of renal ectopias[2]. We are describing a case report of intrathoracic kidney in a paediatric patient.

Case report

A male baby in his second month of life was brought to hospital with a respiratory tract infection of 4 days' duration. The child had noisy breathing and tachypnoea. The weight of the child was 3.85 kg with a birth weight of 2.2 kg, suggestive of failure to thrive. The pulse rate was 146/minute and the respiratory rate was 64/minute. The child had retractions in the subcostal region with deviation of the mediastinum to the right. The breath sounds were well heard bilaterally and were of the vesicular type with rhonchi being present over both lung fields. The heart sounds were prominently heard over the right side of the chest with a systolic murmur best heard in the pulmonary area. Examination of the other systems was normal.

A provisional diagnosis of bronchiolitis was made and treatment was started while awaiting investigations. Chest X ray (Figure 1) showed a white opacity in the right lung with shifting of mediastinum to the right side. The heart was dextroposed. Left lung was normal.

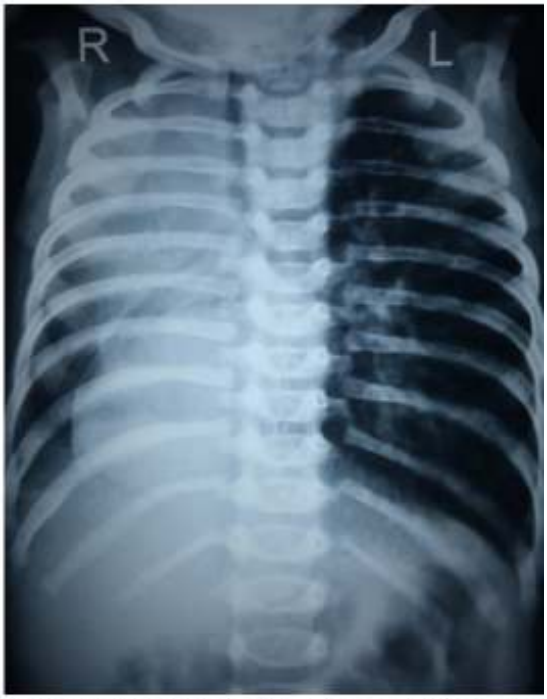

Figure 1: ***Chest x-ray showing white opacity in right lung with shifting of mediastinum to the right side***

2D echocardiogram revealed dextroposed heart with sub-aortic ventricular septal defect (VSD), atrial septal defect (ASD) and severe pulmonary hypertension.

Contrast enhanced computed tomography (CECT) of thorax was suggestive of pulmonary hypoplasia of right upper and middle lobes of lung. The right kidney was herniating through a posterolateral defect in the diaphragm into the thorax. No other bowel loops were seen to be herniating through the defect (Figure 2).

The child was treated for bronchiolitis and recovered well. He is asymptomatic at one year follow up.

[1]*Institute of Medical Sciences and SUM Hospital, Bhubaneswar, Odisha, India*
**Correspondence:*
mohammedwasiq03@gmail.com

https://orcid.org/0000-0002-8920-6819

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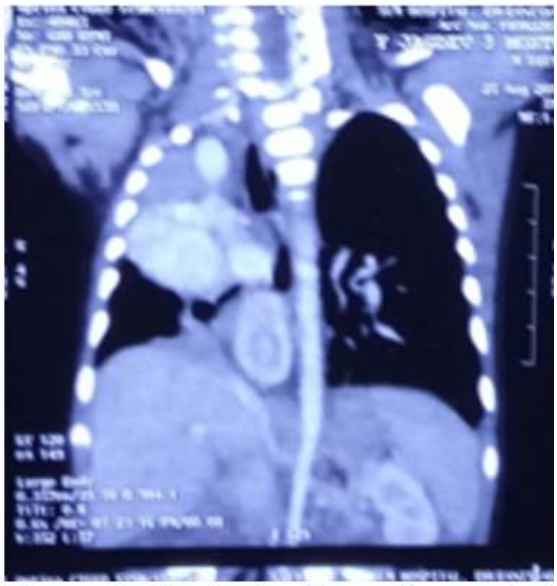

Figure 2: ***Contrast enhanced computed tomography of thorax***

Discussion

The common side of intrathoracic ectopic kidney is the left which is twice as common as the right[3]. In our case the intrathoracic ectopic kidney was on the right side. Intrathoracic kidney is categorized into four groups according to its features[4]:

1. Pure intrathoracic kidney with normal physiology.
2. Intrathoracic kidney with abnormal contour of dome of diaphragm.
3. Intrathoracic kidney with diaphragmatic hernia which is of two types congenital (Type A) or acquired (Type B).
4. Post-traumatic diaphragmatic tear and intrathoracic renal herniation.

The pure intrathoracic kidney is usually not associated with any other malformation. The condition remains asymptomatic and the diagnosis is incidental. Pulmonary symptoms are not so common and urinary symptoms are rarely encountered. The intrathoracic location of a kidney is commonly found to be associated with hypoplasia of the lower lobe of lungs[5]. This is unlike the present case where the hypoplasia is in the upper and middle lobes.

Plain radiographs usually confuse intrathoracic kidney with posterior mediastinal mass (like Bochdalek hernia), neurogenic mass or pulmonary sequestration. A contrast enhanced CT scan can easily diagnose the condition and can also be used as urography to know the functional status of the kidney. Nuclear imaging is very helpful in diagnosing it. Tc 99mDMSA and Tc 99mDTPA scintigraphy are needed to look for the structure and function of the ectopic kidney. However the most cost-effective, non-invasive and reliable method for screening is ultrasonography[6]. The renal vessels and the ureter goes in and out from the pleural cavity through the foramen of Bochdalek respectively.

The other types of intrathoracic kidneys associated with defect in the diaphragm usually have respiratory symptoms and failure to thrive. These children require investigation and further treatment by surgery. As the patient described here had both features of respiratory infection and failure to thrive we referred him to a paediatric surgeon since we suspected a defect in the right diaphragm.

Earlier research had concluded that some predisposing factors like maternal vitamin A or folic acid deficiency, radiation exposure, infectious agents during fetal period, teratogens, trisomy 18 and 21 lead to the development of renal ectopia[7]. The embryogenesis of this anomaly is explained by delayed involution of the mesonephros which leads to more cranial migration of the metanephros[8].

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