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Ectopic thoracic kidney in infancy

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Abstract

Ectopic thoracic kidney is the rarest subtype of ectopic kidney with very few case reports in paediatric age group. We report an interesting case of a 6 months old infant who presented with an episode of pneumonia and was diagnosed to have type 1 thoracic ectopic kidney, which was subsequently confirmed by Computed Tomography (CT) and fluoroscopy. Renal function parameters were normal. Patient was planned for conservative treatment for pneumonia and close follow up with which the child improved. In case of suspected intra-thoracic masses in infants the presence of ectopic kidney should be kept as a differential in order to avoid surprises and indecision on the operating table. Conservative treatment is usually advised in most cases if there is no compression of neighbouring structures or the kidney itself leading to a critical functional compromise.

Keywords

ectopic kidney; thoracic kidney; renal malformations; infancy

Introduction

Ectopic thoracic kidney is a very rare congenital abnormality. Very few cases have been reported of its occurrence in paediatric age group so far. Ectopic thoracic kidney is often detected incidentally during radiography in older age group during imaging performed for evaluation of other diseases or for screening purposes. Most reported cases of thoracic ectopic kidneys in infancy have been commonly associated with Congenital Diaphragmatic Hernia (CDH) and often present with respiratory distress. To the best of our knowledge, there is no report of true left ectopic thoracic kidney without an associated anomaly of the diaphragm presenting with severe respiratory distress in infancy. We are reporting a rare case of ectopic thoracic kidney without diaphragmatic hernia or eventeration presenting with severe pneumonia, early detection of which helped avoid unnecessary intervention.

Case history

A 6 month old male baby, first born child to non-consanguineous parents with no Significant perinatal and developmental history presented with a history of fever, respiratory distress and decreased frequency of urine sine the last 3 days. He was in respiratory distress with saturation of 81% at room air. Heart rate was 178/min, all peripheral pulses were well felt. Complete blood count revealed polymorphonuclear leucocytosis and CRP was positive. Blood urea was 42 mg/dl and serum creatinine

0.4mg/dl. Serum electrolytes were within normal limits. There was no growth of organisms either in blood or urine culture. Chest x-ray was suggestive of elevation of left hemi diaphragm with herniation of lung parenchyma to right side in antero-posterior view (Figure 1A) and the lateral view revealed the possibility of a posterior mediastinal mass. Ultra sound of abdomen showed left kidney present above the spleen. CT thorax showed bulky, slightly malrotated (Figure 1B) Diaphragm was very faintly visualized.

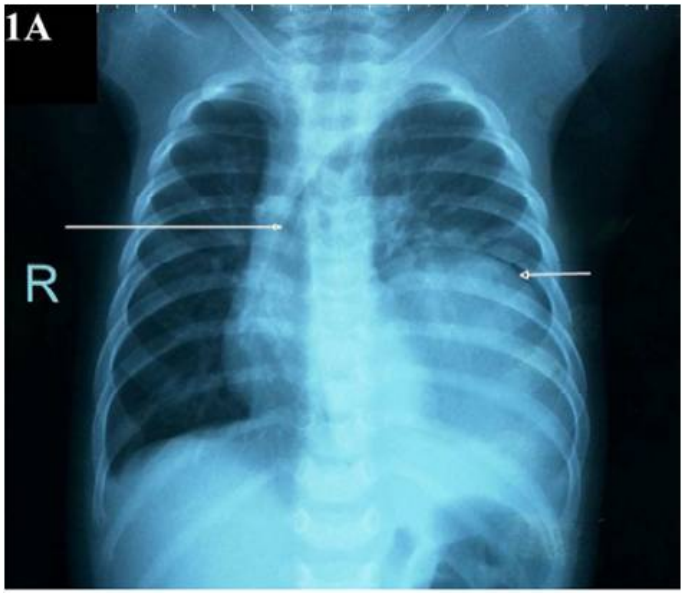

Figure 1A: Apparent elevation of left hemi diaphragm (long arrow) and herniation of left lung (short arrow)

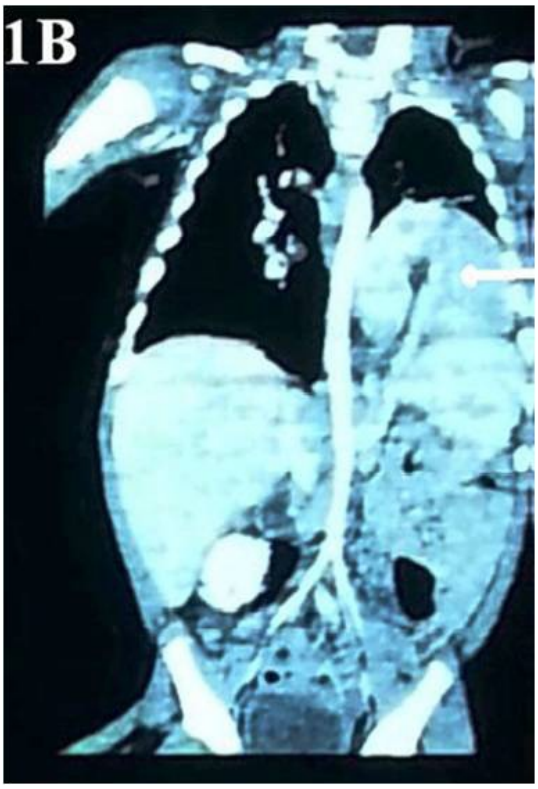

Figure 1B: Ectopic left thoracic kidney in CT chest

He was treated with intravenous amoxicillin-clauvulanic acid and Amikacin for 10 days. Subsequently, Fluoroscopic examination was done which revealed normal position and normal respiratory movement of left hemi diaphragm (figure 2). Both splenic mass and intestinal loops were below the diaphragm. Part of left lung parenchyma which appeared to be herniating to right on the X-ray chest at admission was apparently normal. Right hemi diaphragm was normal in position and had a normal movement with respiratory. Micturating cystourethrogram was performed which was normal. Patient was discharged with advice for follow up. Patient was asymptomatic on latest follow up.

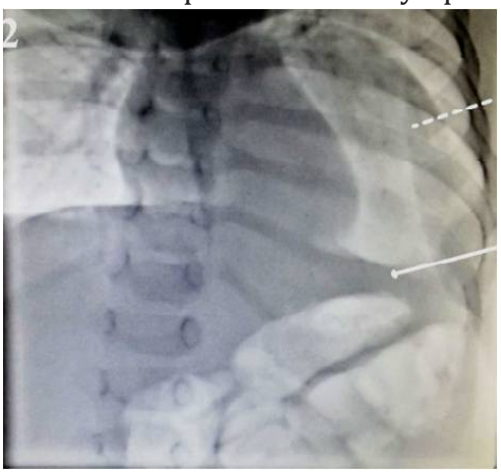

Figure 2: Fluoroscopic demonstration (after treatment) of left hemi diaphragm (arrow) and ectopic thoracic kidney (dotted lines). No herniation of lung parenchyma.

Discussion

Thoracic ectopic kidney is a rare congenital abnormality with an estimated prevalence rate of less than 1 in 10,000. There are few cases which have been reported of its occurrence in paediatric age group so far [1]. Among all types of ectopic kidney the occurrence of the kidney in thorax is the rarest subtype accounting for 5% of all ectopic kidneys [2]. This anomaly often involves either a partial or complete protrusion of the kidney into the posterior mediastinum above the level of the diaphragm. This anomaly is more commonly seen in males and is more common on the left side [3]. In a normal embryo the kidney reaches the adult location by the end of eight weeks of gestation. The embryological defect which is believed to predispose to the occurrence of this anomaly is a defective mechanism causing delay in the closure of the diaphragm in comparison to normal cases or may be related to accelerated ascent of the kidney [4]. Thoracic ectopic kidney have been conventionally classified into 4 types by Pfister-Goedeke L et al [5]. 1. real thoracic ectopic with normally developed closed diaphragm, 2. eventration of diaphragm, 3. diaphragmatic hernia: a) congenital diaphragmatic defects, b) acquired herniation (Bochdalek's), 4. Traumatic rupture of diaphragm with ectopic renal kidney. Based on this classification system our case belongs to type 1, i.e., true thoracic ectopic kidney.

Our case in unique in regards to it being the only reported case of left thoracic ectopic kidney with normal diaphragm to the best of our knowledge, although there has been report in 6 months old infant presenting with respiratory tract infections and right ectopic kidney. In this case thoracic ectopic kidney was suspected based on apparently elevated left hemi diaphragm and herniation of ipsilateral lung parenchyma through mediastinum. The occurrence of an ectopic thoracic kidney is an important cause of apparently elevated hemi diaphragm or mass in chest and usually asymptomatic contrary to its presentation in our case [4]. Thoracic ectopic kidney should be ruled out before any interventions for mass in lower chest or eventeration of diaphragm. This differential should be kept in mind in order to avoid unexpected surprises during improperly planned surgical intervention and also avoid unnecessary investigations. It is also one of the very rare causes of pseudo co-arctation of aorta [6].

Our patient is unique in contrast to the other cases which have been reported to have been diagnosed during infancy (one in antenatal period and 3 during infancy) in regards to the fact that our case had no CDH [7,8]. Our case presented with pneumonia with severe respiratory distress similar to one published by Fouda -Neel K et al [9]. In cases which this or similar anomalies involving the kidney, Tc-99mDTPA scintigraphy can be used for functional evaluation of ectopic kidney. There is report of diagnosing thoracic ectopic kidney by intravenous injection of 5mCi Technetium-99m dimercaptosuccinic acid (Tc- 99m DMSA) scintigraphy when intravenous pyelography failed to visualize the kidney [10].

Conclusion

In case of suspected intra thoracic masses in infants this should be kept as a differential in order to avoid surprises and indecision on the table and inappropriate and potentially life threatening investigations and treatment. Thoracic ectopic kidney should be considered in all patients with apparent elevation of diaphragm in radiography. Suspected cases should be confirmed by computed tomography, scintigraphy, intravenous pyelography and/or fluoroscopy. Patients do not require intervention for renal ectopia, unless they develop a complication. Surgical intervention should preferably be undertaken in

cases with significant functional compromise of the kidney or its neighbouring structures and in cases with relatively low peri-operative risk.

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