Case report

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선천성 횡격막 탈장과 동반된 이소성 흉강내 신장 1례

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Congenital Thoracic Ectopic Kidney associated with Diaphragmatic Hernia in a 15-month-old Boy

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Congenital thoracic ectopic kidney is a very rare developmental disorder and the rarest type of ectopic kidney. This condition is usually asymptomatic and detected incidentally on routine chest radiography. Most cases of thoracic ectopic kidney develop in adulthood and during the neonatal period, and congenital thoracic ectopic kidney rarely develops in children. Most patients are asymptomatic, and the treatment depends on the diagnosis. Herein, we report a rare case of ectopic thoracic kidney associated with a diaphragmatic hernia in a 15-month-old male infant, who presented with periodic severe irritability. The thoracic ectopic kidney was detected as a mass in the right base of the chest on routine chest radiography.

Key words: Intrathoracic kidney, Diaphragmatic hernia

Introduction

Congenital thoracic ectopic kidney is a very rare condition, with one study showing it represented less than 5% of all ectopic kidneys in a series of 13,000 autopsies [1]. A thoracic ectopic kidney may be caused by a congenital anomaly or may occur secondary to diaphragmatic hernia. The incidence of an ectopic kidney is higher on the left side and in males. Diagnosis is usually incidental, and it may be detected on routine chest radiography as an elevated hemidiaphragm. To diagnose a thoracic ectopic kidney, computed tomography (CT) or pyelography may be useful. Most patients are asymptomatic, and the treatment depends on the diagnosis. In this case report, we describe a male infant with thoracic ectopic right kidney and a congenital diaphragmatic hernia.

Case report

A 15-month-old boy with a history of periodic severe crying when lying down in the supine position was referred to our hospital for further evaluation. He had been delivered by caesarian section at 40 weeks, weighing 2.8 kg (10-25th percentile), and had an uneventful birth. No unusual urologic history was reported.

The infant weighed 10.0 kg (25th percentile) and was 82.4 cm (50-75th percentile) in length on admission. Physical examination revealed a diminished right-sided breath sound, but no anomaly of external genitalia. The infant was otherwise healthy, with normal development for his age.

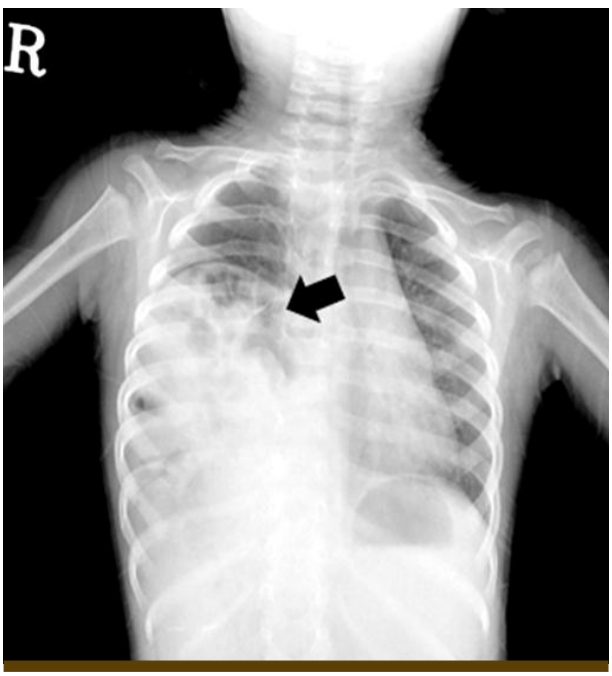

Fig. 1. Chest radiograph (posterior-anterior view) showing normal small bowel gas (arrow) with elevation of the right diaphragm.

Laboratory studies were done at admission. Findings of urinalysis were normal. Complete blood count was performed; total white blood cell count was 9,800/mm^3 (normal range: 6,000-15,000/mm^3), with neutrophils at 35.8% (normal range: 40-80%). Serum electrolytes were within the normal range.

A chest radiograph showed a decreased volume of the right lung and a normal bowel gas pattern below an elevated right diaphragm (Fig. 1). The patient underwent renal ultrasonography and 99-technetium dimercaptosuccinic acid (DMSA) scintigraphy. On ultrasonography, the left kidney showed normal echogenicity with compensatory hypertrophy, but the right kidney was not detected (Fig. 2). DMSA scintigraphy diagnosed an ectopic right kidney in the superior thoracic cavity (Fig. 3). Magnetic resonance imaging (MRI) revealed a defect in the right diaphragm and herniation of the small and large intestine on the right side of the body (Fig. 4).

During corrective surgery, a defect in the diaphragm was found on the right posterior side. The right kidney, entire small intestine, and ascending colon were located in the thorax, without strangulation. No other organic anomaly was seen, except for cecal malrotation. The right kidney was correctly positioned into the abdominal cavity, and the herniated small intestine and ascending

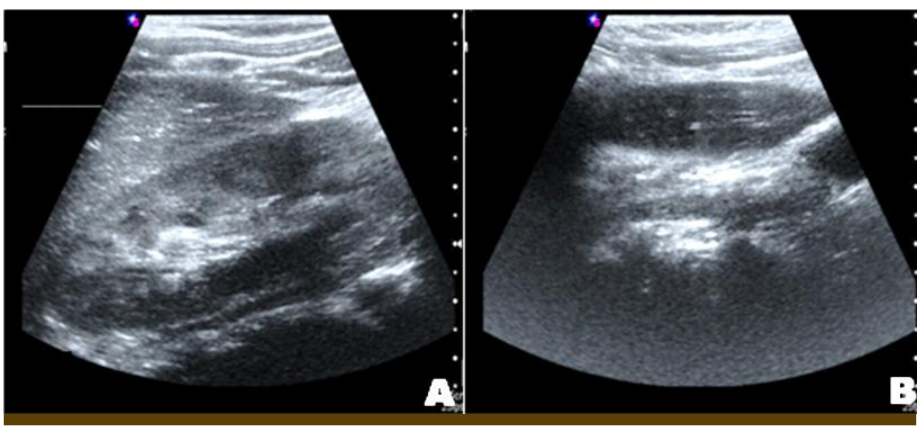

Fig. 2. Ultrasonography showing normal left kidney measuring 7.3 cm (A) and absence of the right kidney in the renal space (B).

colon were also extracted. The diaphragm was repaired using polytetrafluoroethylene (PTFE) patch (Gore tex®: W.L. Gore & Associates) to close the defect. The cecum was relocated by ligation to the base of the appendix, and appendectomy was performed.

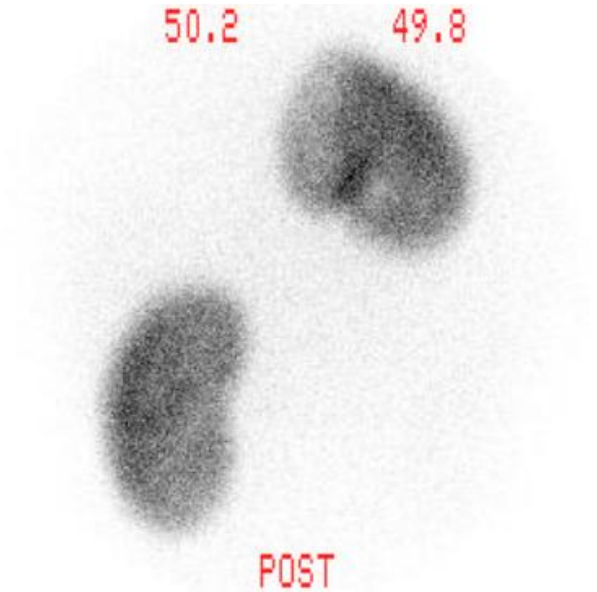

Fig. 3. 99-technetium dimercaptosuccinic acid (DMSA) scintigraphy showing the high positioned kidney of right with the hilum pointing downward. POST: posterior view.

Postoperative intravenous pyelography (IVP) performed 1 week after surgery showed that both kidneys were located in the intra-abdominal cavity with normal pelvocalyceal system appearance (Fig. 5). The child had an uneventful postoperative course and was doing well at follow-up three years after surgery.

Discussion

Ectopic kidneys occur in only one in 10,000 births and are found in pelvic, iliac, abdominal, or thoracic locations. Congenital thoracic ectopic kidneys account for less than 5% of ectopic kidneys [1, 2]. Of the 22 ectopic kidneys detected in an autopsy series of 15,919 pediatric patients, only one case of an intrathoracic kidney was reported [3]. The first report of a thoracic ectopic kidney was in a 43-year-old woman in 1940 [4]. Thoracic ectopic kidneys present most often on the left side (in a ratio of 1.5:1) and are twice as common in males [5, 6]. Unlike pelvic ectopic kidneys, thoracic ectopic kidneys can cause respiratory difficulty or be asymptomatic [7, 8]. In most asymptomatic cases, thoracic ectopic kidneys are incidental findings on routine

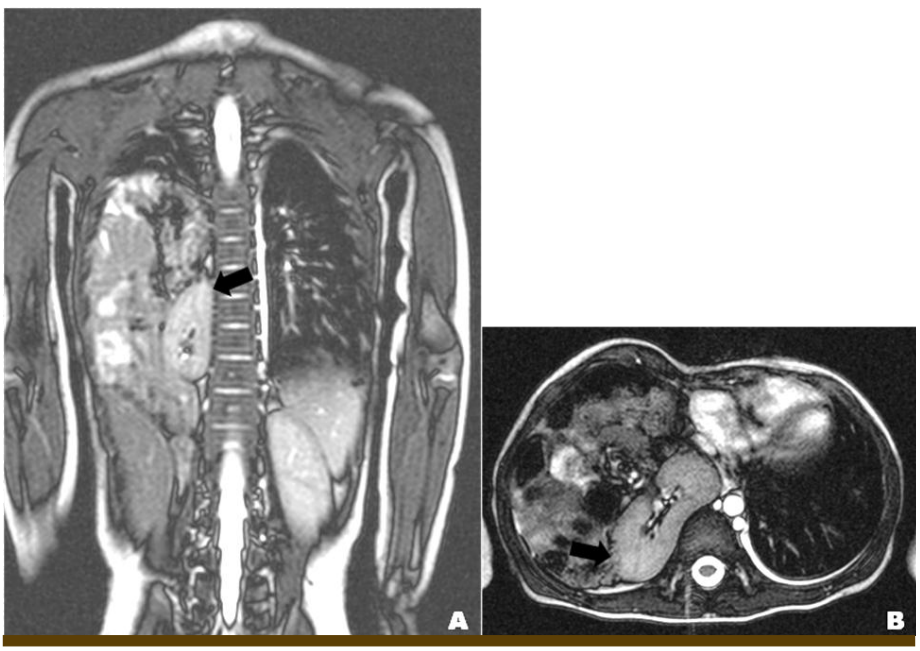

Fig. 4. Magnetic resonance imaging showing coronal (A) and axial (B) views with the right kidney (arrow) and the small and large intestines protruding above the diaphragm on the same side.

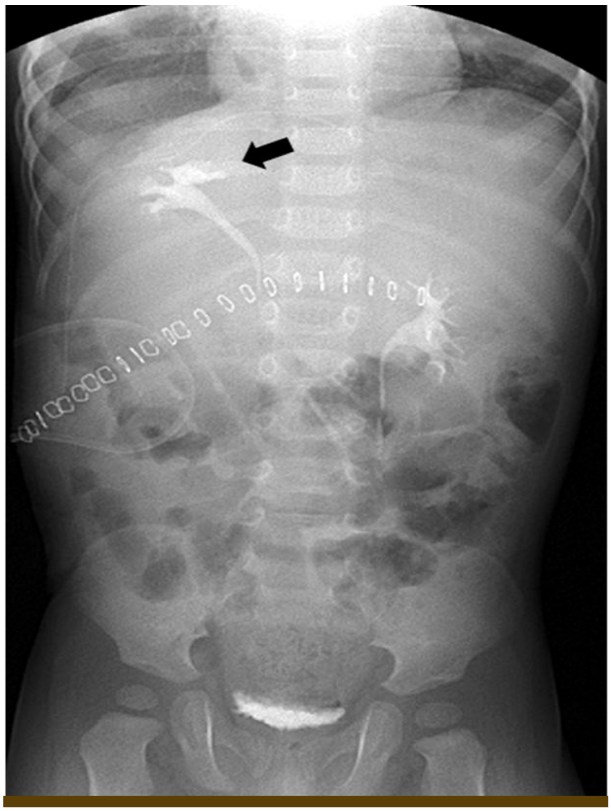

Fig. 5. Postoperative intravenous pyelography showing correctly positioned ectopic right kidney (arrow) in the abdominal cavity with normal appearance of the pelvocalyceal system.

chest radiography [6].

Congenital thoracic ectopic kidney can be defined as an excessive cranial ascent of the kidney which is usually completed by the 8th week of gestation [9]. Accelerated ascent of the kidney preceding to closure of the pleuroperitoneal membrane is possibly thought to be a mechanism leading to a congenital thoracic ectopic kidney [10]. And mal-development or delayed closure of the pleuroperitoneal membrane may be other possible causes [10, 11]. The early closure of the right sided pleuroperitoneal membrane and the presence of the liver limit the incidence of a thoracic ectopic right kidney, but it can occur because of a right-sided diaphragmatic hernia [12]. The other suggested mechanisms of congenital thoracic ectopic kidney have an effect on the developing liver or adrenal gland on renal position, and congenital intrinsic factors in the nephrogenic system. However, none of these mechanisms can be adequately explained the role in the development of a congenital thoracic ectopic kidney.

The incidence of intrathoracic kidneys associated with congenital diaphragmatic hernia is less than 0.25% [13]. Ptister-Goedeke and Brunier described four types of thoracic ectopic kidneys: (1) thoracic renal ectopia with a normal diaphragm; (2) eventurated diaphragm; (3) diaphragmatic hernia (congenital or acquired); and (4) traumatic rupture of the diaphragm with renal ectopia [14-16]. Our case corresponds to that of thoracic ectopic kidney with diaphragmatic hernia.

The congenital thoracic ectopic kidney is mostly independent abnormality. Included our patient, most cases are seen the adrenal gland below the kidney. In spite of associated abnormalities are rare [9], there are reported some cases with an associated colonic malposition in our case or ectopic adrenal gland or spleen. Lundius B et al reports the case of bilateral thoracic ectopic in a 63-year-old man. And one case of a thoracic kidney with an congenital heart disease, acyanotic tetralogy of Fallot is described in a 5-year-old boy. Then, we carefully suggest that evaluation of associated abnormalies in patient with ectopic kidney should be done.

The radiographic appearance of congenital thoracic ectopic kidney may be similar to that of other thoracic masses such as Bochdalek hernia, sequestration, and neurogenic masses. In some cases, the only finding is elevation of the diaphragm [15]. Because the herniated ectopic kidney is usually positioned in the posterior mediastinum, it is highly suggested to the reniform mass. Therefore, computed tomography (CT) helps not only the detection of mediastinal mass but also the confirm the diagnosis with pelvocalyceal structures and soft tissue density of the kidney. And DMSA scintigraphy provides a simple and non-invasive evaluation of the location, size, and shape of the kidney and can be used to investigate renal function and cortical uptake [17].

A congenital thoracic ectopic kidney does not require treatment, but a thoracic ectopic kidney associated with congenital hernia or traumatic rupture of the diaphragm needs to be treated with surgery [18, 19]. In addition, a thoracic ectopic kidney associated with hematuria, stone formation, ureteropelvic tract obstruction, or flank pain must be considered for specific interventions [20].

The present case showed a right-sided congenital diaphragmatic hernia. No history of trauma was reported; therefore, the cause involved ascent of the intestine and right kidney. The case was managed with surgery.

In conclusion, congenital thoracic ectopic kidney with diaphragmatic hernia is a rare condition but should considered in the presence of abnormal findings on chest radiography in a patient with positional respiratory symptoms. CT or MRI can be used to diagnose a congenital thoracic ectopic kidney associated with diaphragmatic hernia.

Acknowledgment

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한글요약

선천성 흉강내 신장은 이소성 신장의 가장 드문 형태로 알려져 있다. 흔히 증상이 없어 우연히 흉부 방사선 촬영을 통해 발견 되며, 주로 성인 시기와 산전 진단에서 진단 받는 경우가 많은 것으로 알려져 있다. 선천성 흉강내 신장은 다른 동반기형이 없는 무증상의 경우 수술적 치료가 필요하지 않으며, 횡격막 탈장이 동반된 경우 출생 후 수일 이내에 교정 수술이 필요 하다. 이에 저자들은 분만력, 과거력상 특이 소견이 없었던 15개월 남아로 간헐적인 심한 보챔을 주소로 내원하였고 흉부 방사선 촬영에서 종격동 종괴를 보여 시행한 초음파와 스캔에서 신장 위치의 이상을 확인하였고, MRI를 통해 횡격막 탈장을 확인한 예를 경험하였기에 보고하는 바이다.

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