

Ectopic Thoracic Kidney in a Child with Congenital Diaphragmatic Hernia

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Abstract

Thoracic ectopic kidney with partial or complete renal protrusion above the level of the diaphragm into the posterior mediastinum is the rarest form of all ectopic kidneys with an incidence of less than 1 per 10 000 cases. We present a newborn with right congenital diaphragmatic hernia associated with thoracic ectopic kidney. The diagnosis of ectopia was made prior to surgery. Gerota's fascia of kidney was used to close the diaphragmatic defect. Since this renal anomaly is usually asymptomatic, it does not require any specific treatment. However, a close examination of function and anatomy of the kidney prior to surgery of hernia is important and beneficial. We discuss the embryological context and the importance of renal scintigraphy in patients with ectopic kidney.

Key words

Intrathoracic kidney

Résumé

L'ectopie rénale intra thoracique partielle ou complète avec ascension du rein au-dessus du niveau du diaphragme dans le médiastin postérieur est la plus rare de toutes les ectopies rénales, avec une incidence de moins de 1 sur 10 000 cas. Nous présentons le cas d'un nouveau-né avec une hernie diaphragmatique droite associée à une ectopie rénale. Le diagnostic de l'ectopie a été réalisé avant la chirurgie. Le fascia de Gerota est utilisé pour fermer le défaut diaphragmatique. Bien que cette anomalie rénale soit habituellement asymptomatique, elle ne nécessite au-

cun traitement spécifique. Néanmoins, apprécier la fonction et l'anatomie du rein avant la chirurgie est important et bénéfique. Nous discutons l'aspect embryologique de cette association et l'importance de la scintigraphie rénale chez les patients avec une ectopie rénale.

Mots-clés

Rein intrathoracique

Resumen

El riñón torácico ectópico, la protrusión parcial o completa del riñón por encima del nivel del diafragma en el mediastino posterior, es la forma más rara de ectopia renal con una incidencia menor de 1 por 10 000 casos. Presentamos un recién nacido con una hernia diafragmática congénita derecha asociada con un riñón ectópico torácico. Se hizo el diagnóstico de la ectopia antes de la cirugía. La fascia de Gerota del riñón fue usada para cerrar el defecto diafragmático ya que ésta anomalía renal es generalmente asintomática y no requiere ningún tratamiento específico. Sin embargo, es útil y beneficioso evaluar la anatomía y la función del riñón antes de la cirugía de la hernia. Discutimos una revisión embriológica de esta asociación y la importancia de la escintigrafía renal en pacientes con riñón ectópico.

Palabras clave

Riñón intratorácico

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Zusammenfassung

Die seltenste Form aller ektopten Nieren ist die partielle oder vollständige Verlagerung in den Thorax über das Zwerchfell. Ihre Häufigkeit beträgt weniger als 1 : 10 000 Fälle. In der vorliegenden Arbeit wird ein Neugeborenes mit einer intrathorakalen Nierenektomie vorgestellt. Um die gleichzeitig bestehende Zwerchfelllücke zu schließen, wurde die Gerotasche Faszie verwandt.

Introduction

Ectopic positioning is not the rarest anomaly of the kidney. However, a thoracic location of the ectopic kidney is most unusual. Only 22 ectopic kidneys were found out of a total of nearly 16000 autopsies performed in a series, and only one of these was intrathoracic [3]. Thoracic ectopia of the kidney may be a congenital anomaly or secondary to herniation through a congenital or acquired diaphragmatic defect; males are affected preponderantly and the left side is more commonly involved [2].

The discovery of this rare association prior to surgery is important for the management in diaphragmatic hernia repair. It is sometimes not easy to create a new diaphragm, especially in cases with transdiaphragmatic thoracic kidneys. We also wish to emphasize the importance of scintigraphic studies in these cases.

Case Report

A 10-day-old baby girl was admitted to the Children's Clinic with the complaints of cyanosis and hypotonia. Prenatal, natal, and postnatal history were unremarkable. The physical examination revealed respiratory distress and no lung voices could be heard in the right hemi-thorax. Other systems showed no pathological findings.

Chest X-ray revealed a well delineated, right congenital diaphragmatic hernia. Abdominal ultrasonography showed that the right kidney was located superior to its normal location. After this finding, we found necessary to examine the location of the right kidney in detail prior to surgery. Computed tomography (CT) revealed posterolateral herniation of the diaphragm and renal thoracic ectopia on the same side (Fig. 1).

During the operation, a thoracic ectopic right kidney was found at the level of T7–8 for the upper pole. All contents of the hernia were relocated to the abdominal cavity except for the kidney. The kidney was fixed to the retroperitoneum and we were unable to withdraw it. No gross developmental anomaly was seen during the operation except for renal malrotation. We did not find the posterior leaf of the diaphragm sufficiently developed for closure of the defect. The diaphragmatic leaf was therefore sutured to Gerota's fascia for closure of the defect.

Präoperativ wurde die Nierenfunktion und Anatomie geklärt, was auch postoperativ nach Verschluss der Zwerchfellhernie zwingend notwendig ist. In der Arbeit werden die embryonalen Zusammenhänge, die Bedeutung der Nierenzintigraphie bei ektopten Nieren diskutiert.

Schlüsselwörter

Intrathorakale Nierenlage · ektope Niere

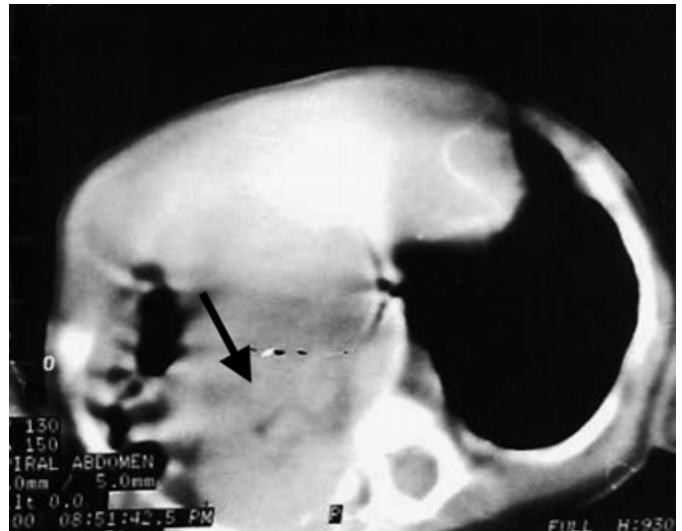


Fig. 1 Enhanced thoraco-abdominal computed tomography reveals right kidney's intrathoracic location. Arrow indicates the right kidney.

In the postoperative period, renal cortical scintigraphy with Tc-99 m DMSA (dimercaptosuccinic acid) was performed for evaluation of the relative function of the kidneys (Fig. 2). After 3 hours' intravenous injection of 0.5 mCi (18.5 MBq) Tc-99 m DMSA, planar images (in six positions) of the kidneys were obtained using a single head gamma camera. According to the images, the left kidney was in its normal position and had a normal shape and size. However, the right kidney was in the thoracic region. Visually both kidneys had an equal relative functional uptake and no cortical defect was detected. In order to localize the ectopic kidney precisely and to evaluate its relationship to the lungs and lung functions, pulmonary perfusion scintigraphy was performed soon after DMSA imaging (Fig. 2).

Six months after surgery, the child was again admitted to our department for dyspnea. Chest X-ray revealed diaphragmatic eventration. We also examined the diaphragm thoracoscopically because of a suspicion of recurrence of the diaphragmatic herniation as we had used Gerota's fascia for the closure. No diaphragmatic herniation was found (Fig. 3). We performed plication of the diaphragm with thoracotomy. The appearance of the kidney was normal except for its malrotation. It was found to be more mobile than after the first operation and was withdrawn to the abdominal cavity. The child has been followed up now for 2 years without complaints and no renal pathology was detected with scintigraphic studies.

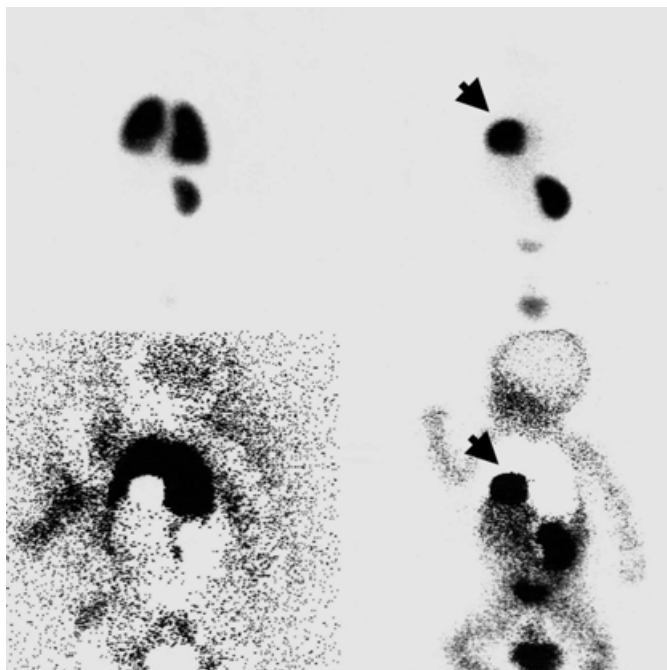


Fig. 2 Tc-99m DMSA images in the anterior position (top right). The left kidney in a normal location and the ectopic right kidney (arrow) in the thoracic cavity showed normal cortical uptake of Tc-99m DMSA. It was suggested that the ectopic kidney had a rotational anomaly since the inferomedial portion had a slightly lower uptake. Pulmonary perfusion and DMSA images in the posterior position (top left), subtraction images in the anterior position (bottom) demonstrated the exact location of the ectopic kidney, the relationship between the ectopic kidney and the right lung, and the function of lungs. Normal perfusion was shown in both lungs except in the right lung, in the region of the ectopic kidney.

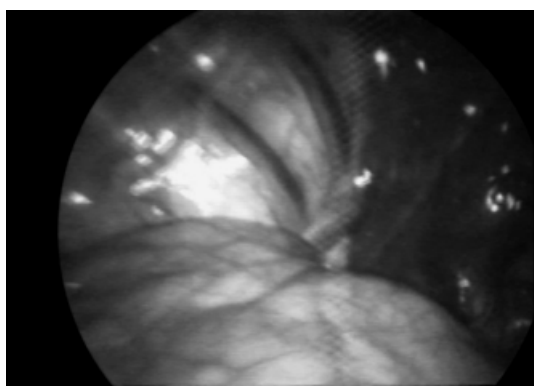


Fig. 3 Thoracoscopic view of the right thoracic cavity. No reherniation was detected.

Discussion

Ectopic thoracic kidney can be simply defined as an excessive cranial ascent of the kidney which is normally completed by the 8th week of gestation [7]. Initially, an accelerated ascent of the kidney prior to diaphragmatic closure was thought to be a possible mechanism leading to a thoracic kidney. Delayed closure or maldevelopment of the pleuroperitoneal membrane that allows continued ascent were other possible mechanisms thought to be responsible for an intrathoracic kidney [2]. However, this is now considered to be unlikely since the incidence of intrathoracic

kidney as an associated anomaly with congenital diaphragmatic hernia has been found to be less than 0.25 percent [3].

The other suggested possible mechanisms include the influence of the developing adrenal gland and liver on renal position and intrinsic factors in the kidney such as the hypothesis that a congenital thoracic kidney is secondary to persistence of the nephrogenic cord [2]. A case of right intrathoracic kidney with complex axial skeleton defects which included vertebral fusion and right intrathoracic supernumerary ribs was published and this association of a congenital intrathoracic kidney tends to favor the last theory [1]. However, none of these mechanisms can be fully excluded and each may have a role in the development of a thoracic ectopic kidney.

The location of a thoracic ectopic kidney may be supra, infra, or trans diaphragmatic [7]. A normal kidney's superior border is positioned between T10–L1. If the upper border of the thoracic kidney is between T7–11, this is referred to as transdiaphragmatic ectopia. In our case, the upper pole of the kidney was at the T7–8 level and was therefore a transdiaphragmatic ectopia. The thoracic kidney in a diaphragmatic hernia is mostly mobile and can be easily withdrawn from the thorax [5]. In our case it was fixed to the retroperitoneum and we were unable to withdraw it during the first operation but succeeded in the second.

In some conditions it is not possible to close a diaphragmatic defect, and rib or abdominal muscle flaps, or prosthetic meshes can be used to close the defect. In our case, the location and fixation of the kidney did not permit us to create a new diaphragm easily. We therefore sutured Gerota's fascia to the diaphragmatic edge. As far as we are aware, there is no published report on diaphragmatic hernia repair using Gerota's fascia for closure. If we could have repositioned the ectopic kidney in the abdominal cavity during the first operation, it is possible that eventration could have been avoided and the diaphragm could have been closed with or without a patch in a conventional manner. The eventration that happened in this case is important as it may have been due to a relaxation of Gerota's fascia. However, in the second operation we did not find any opening which would have permitted reherniation or any direct relaxation of Gerota's fascia. But there is no doubt that more experience is necessary before any conclusion can be drawn concerning the use of Gerota's fascia in diaphragmatic hernia repair.

Although associated anomalies in other organ systems are rare, cases have been reported with an accompanying ectopic adrenal gland, ureteropelvic junction obstruction and malpositioning of the affected kidney [4, 7]. Invariably, malrotations of the kidneys are probably due to the fact that the ascending kidney undergoes a ninety degree rotation along its axis. In our case we also found a rotation anomaly which was not associated with any other anatomical defect. This was best demonstrated with scintigraphic studies. We therefore suggest that DMSA renal scintigraphy must be performed in these cases, even if the CT is normal in order to define associated renal anomalies.

Renal cortical scintigraphy with Tc-99m DMSA is known to be a simple and non-invasive method for evaluation of the shape, size, and location of the kidneys (including ectopic kidneys),

and the functional state of renal parenchyma, and for determination of cortical uptake and relative renal functions [6]. It has a low radiation dose and no side effects. After intravenous injection, Tc-99 m DMSA is taken up by tubular cells of the pars recta [8,9]. Since the parenchyma is visualized without interference from the pelvicalyceal system with low background activity (activity in the vascular space or other organs other than kidneys), good images and a good estimation of the relative function can be obtained [8]. Since in our case viability and functional status of the ectopic kidney was very important in the postoperative period, we performed renal cortical scintigraphy with Tc-99 m DMSA. Nevertheless, we will carry out scintigraphic controls periodically to check out the functional status.

In conclusion, thoracic ectopic kidney with Bochdalek's hernia is a rare association and must be defined properly prior to surgery to ensure the best outcome during the operation. No treatment for the ectopic position of the kidney is necessary once the diagnosis of intrathoracic kidney without an associated anomaly has been confirmed. However, close follow-up, especially with scintigraphy, is important for such cases.

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