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CASE REPORT

Bochdaleck hernia and pulmonary sequestration: case report of an unusual association

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Abstract

Introduction: Although bochdalek hernia is the most common type of congenital diaphragmatic hernia, it is still a rare condition. It can be complicated by the development of secondary pulmonary sequestration, which is subdivided into intralobar pulmonary sequestration and extralobar pulmonary sequestration. **Case report:** This study describes a case of pulmonary sequestration due to bochdalek hernia in a one-year-old girl, who underwent surgical correction. **Discussion:** Pulmonary sequestration can be confused with other more frequent pathologies, such as pneumonia or tumors, and diagnosis can be facilitated through the use of vascular imaging techniques, which confirm the diagnosis and show the vascularization of the hernia content.

Keywords: Congenital diaphragmatic hernia. Bochdalek hernia. Bronchopulmonary sequestration. Case report

Hérnia de Bochdalek e sequestro pulmonar: relato de caso de uma associação incomum

Resumo

Introdução: A hérnia de Bochdalek representa a causa mais comum de hérnia diafragmática congênita e ainda é uma condição rara. Pode ser complicada pelo desenvolvimento de sequestro pulmonar secundário, que se subdivide em sequestro pulmonar intralobar e sequestro pulmonar extralobar. **Relato de caso:** Este estudo descreve um caso de sequestro pulmonar devido a hérnia de Bochdalek em uma menina de 1 ano de idade, submetida à correção cirúrgica. **Discussão:** O sequestro pulmonar pode ser confundido com outras patologias mais frequentes, como pneumonia ou tumores, e o seu diagnóstico pode ser facilitado através da utilização de exames de imagem vascular, que confirmam o diagnóstico e mostram a vascularização do conteúdo herniário.

Palavras-chave: Hérnias diafragmáticas congênitas. Hérnia de Bochdalek. Sequestro broncopulmonar. Relato de caso.

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Keypoints

What is known
- Although rare, bochdalek hernia is the most common type of congenital diaphragmatic hernia.
- We need to be aware that congenital pulmonary abnormalities can be associated with congenital diaphragmatic hernia.

What is added
- Lobectomy or, in selected cases, segmentectomy is the treatment of choice for symptomatic and asymptomatic patients with ILPS.
- Thoracoscopy versus thoracotomy in the pediatric population showed no difference in outcomes.
- If the patient remains asymptomatic for a long time, the plan can be changed from surgery to follow-up.

Introduction

Bochdalek hernia (BH) is the most common type of congenital diaphragmatic hernia (CDH), although it is still a rare condition occurring in 1:2,500 live births[1]. A posterior defect in the pleuroperitoneal compartment allows herniation of abdominal viscera into the thorax[1], such as the stomach, omentum, liver, spleen, and intestines. Neonate liver hernia has a poorer prognosis out of all BH[2].

About 50-60% of affected individuals have isolated CDH; the remainder have complex CDH, that is, CDH occurring with additional malformations[3]. One of the more common pulmonary anomalies found with CDH is extralobar bronchopulmonary sequestration (BPS)[4]. More than 50% of cases with CDH are detected prenatally through ultrasound examination[3].

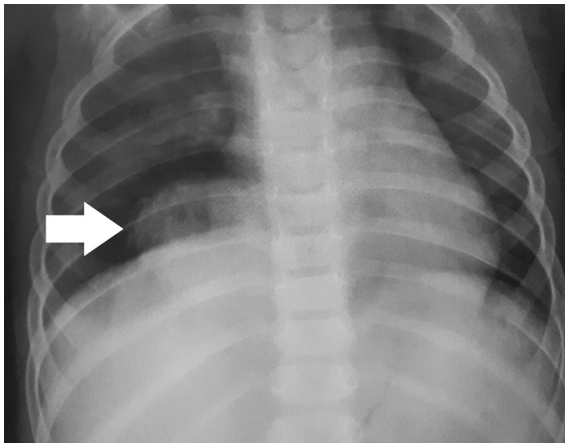

Figure 1. Chest X-ray in a posteroanterior view showing opacity in the right lung base (white arrow).

Case report

A one-year-old girl was seen at the pulmonology department to evaluate an opacity in the right lung base on a chest X-ray (Fig. 1) performed four months after treatment for bacterial pneumonia. The patient had no related symptoms and presented with normal physical examination. Her mother reported normal prenatal examinations and no postnatal complications.

A chest CT scan was performed, which detected opacity in the right lung base with anomalous vascularization from the descending aorta with Bochdaleck diaphragmatic herniation of hepatic content. MRI confirmed right Bochdaleck diaphragmatic herniation with hepatic content (Fig. 2) in addition to associated pulmonary intralobar sequestration (Fig. 3). The patient was referred for surgical treatment, but due to the pandemic the surgery was postponed for two years. During these years the patient remained asymptomatic. Thus, the plan was changed from surgery to follow-up.

Discussion

Pulmonary sequestration (PS) is a congenital malformation consequence of nonfunctioning lung tissue with no tracheobronchial tree communication. PS accounts for 0.15% and 6.5% of all congenital pulmonary malformations[5] and is subdivided into intralobar PS (ILPS) and extralobar PS (ELPS). ILPS has a higher incidence (about 80% of all PS) and is characterized by a blend of the hernia with the normal lung, whereas ELPS is separated from the normal lung by its distinct visceral pleura[6]. The sequestrated lung is supplied by an anomalous artery arising from the aorta and its venous drainage is via the azygous system, the pulmonary veins, or the inferior vena cava. In some cases, the mass effect sequestration is speculated to have a protective effect in concomitant CDH, delaying the herniation of abdominal contents until after delivery and allowing prenatal lung development[3].

Lobectomy or, in selected cases, segmentectomy is the treatment of choice for symptomatic and asymptomatic patients with ILPS, through thoracotomy or thoracoscopy. Recent reviews of thoracoscopy versus thoracotomy in the pediatric population showed no difference in outcomes[7].

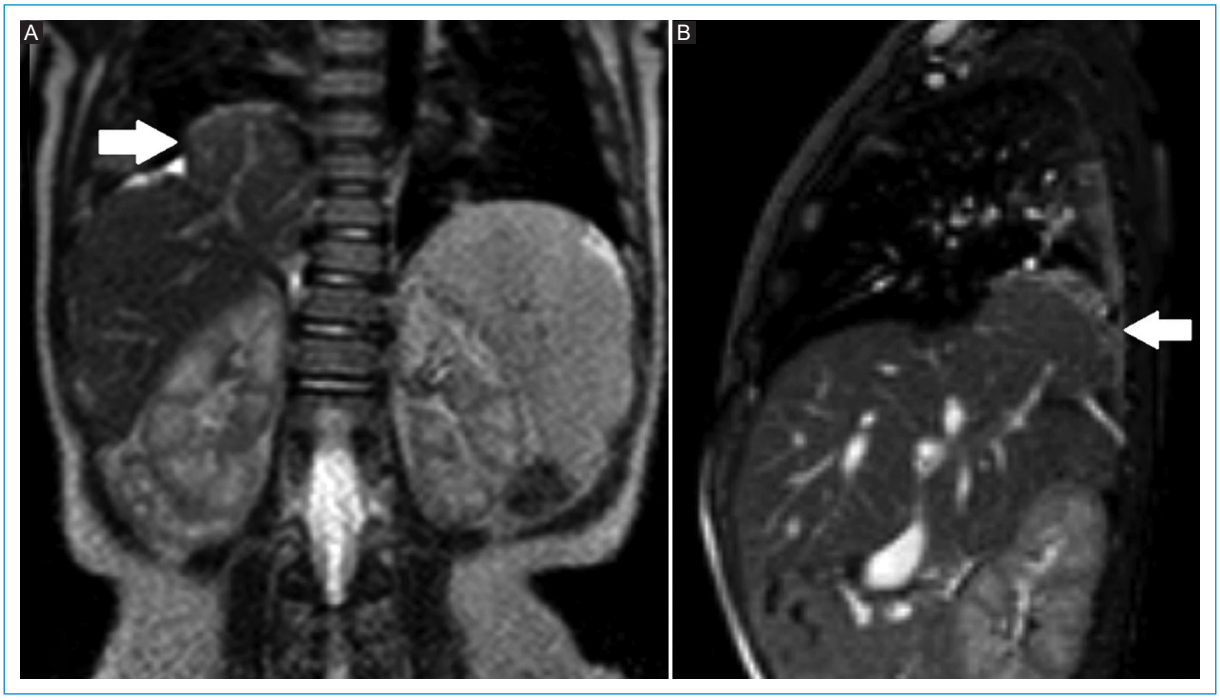

Figure 2. Magnetic resonance in the T2 sequence in **A:** the coronal plane and **B:** sagittal showing Bochdaleck diaphragmatic herniation of hepatic content (white arrows).

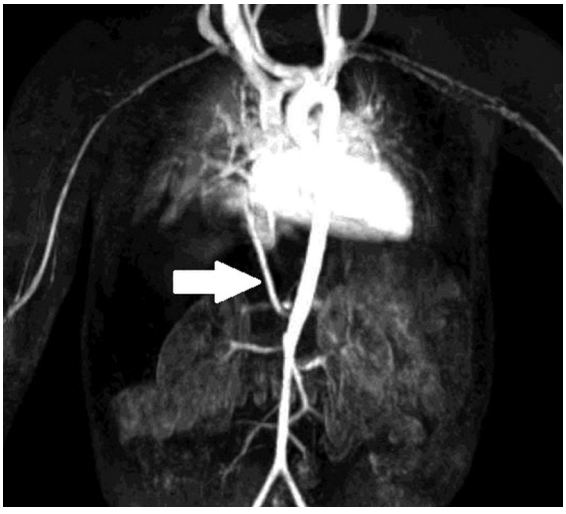

Figure 3. 3D arterial magnetic resonance angiography in the coronal plane showing the arterial vessel originating from the descending aorta supplying the lower lobe of the right lung (white arrow).

The Ramsport et al. systematic review reported that 86% of cases of Bochdalek hernia are treated surgically[8]. All of the patients treated conservatively were asymptomatic[8]. Laparotomy and laparoscopy are the preferred surgical procedures, followed by thoracotomy, thoracoabdominal approach, and thoracoscopy[8]. Some of these procedures are already being performed by robotic surgery[8]. The most common complications of surgical treatment are lung abscess, empyema, pleural effusion, broncho-pleuro-colonic fistula, hemothorax, and pneumonia[8].

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