Successful management of congenital diaphragmatic eventration in a neonate. - Case Report

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Abstract

Congenital diaphragmatic eventration is a rare condition characterized by abnormal elevation of the diaphragmatic muscle, leading to respiratory dysfunction. Its incidence, although low, represents a significant clinical challenge due to the variety of manifestations and the need for a multidisciplinary approach. The use of advanced diagnostic tools, such as radiographs and ultrasound, highlights the crucial role of technology in the early identification of this condition. Specifically, surgery emerges as an essential measure, especially in low birth weight neonates, where it becomes the most effective therapeutic option to prevent serious complications and favor proper lung development. The successful application of minimally invasive surgical techniques highlights the importance of innovation in the management of complex cases.

Resumen

La eventración diafragmática congénita es una condición poco común caracterizada por la elevación anormal del músculo diafragmático, lo que

conlleva disfunción respiratoria. Su incidencia, aunque baja, representa un desafío clínico significativo debido a la variedad de manifestaciones y la necesidad de un enfoque multidisciplinario. La utilización de herramientas de diagnóstico avanzadas, como radiografías y ecografías, destaca el papel crucial de la tecnología en la identificación precoz de esta afección. Específicamente, la cirugía emerge como una medida esencial, especialmente en neonatos con bajo peso al nacer, donde se convierte en la opción terapéutica más efectiva para prevenir complicaciones graves y favorecer el desarrollo pulmonar adecuado. La aplicación exitosa de técnicas quirúrgicas mínimamente invasivas resalta la importancia de la innovación en el manejo de casos complejos.

Introduction

Diaphragmatic eventration (DE) is defined as an uncommon pathology characterized by abnormal elevation of the diaphragmatic muscle, with paradoxical mobility, resulting from a failure in the migration of myoblasts towards the pleuroperitoneal membrane or a deficiency in innervation. This anomaly can be total or partial and tends to be located mainly in the left hemidiaphragm, leading to respiratory dysfunction (1).

With a reported incidence of 0.05 per 1,000 live newborns, DE represents 5% of all diaphragmatic pathologies (2). This condition most frequently affects males and tends to manifest predominantly unilaterally. In congenital or idiopathic cases, there is a slight predominance on the right side, while in acquired cases, the predominance leans toward the left side (3).

Although diaphragmatic eventration may be symptomless, early diagnosis is possible if signs such as tachypnea, dyspnea, recurrent pneumonias or pulmonary atelectasis are present. In addition to these symptoms, less common manifestations such as emesis and fatigue during feeding may occur. Although rare, vascular dysfunction and cardiac manifestations may also occur. Severe respiratory distress is uncommon in most cases (4). Diagnosis is usually established by radiological findings, this being the main diagnostic tool (3).

The treatment of this pathology in symptomatic patients is by surgical procedure. However, the decision to perform surgery in asymptomatic patients remains controversial. Surgical intervention is generally recommended when the diagnosis is made during infancy, especially if the diaphragmatic base is significantly elevated. This is done in order to prevent hypoplasia of the ipsilateral lung and ensure its adequate development (5).

Considering the low incidence both internationally and nationally, this article aims to show the successful surgical management of a premature newborn with the right diaphragmatic eventration.

Clinical case

Preterm female newborn (29.1 weeks), with adequate birth weight, mother 32 years old, secundigestant. Adequate prenatal controls without anomalies.

She was born after emergency cesarean section due to severe oligohydramnios secondary to premature rupture of membranes (18 hours). After birth, she showed signs of severe respiratory distress at birth requiring positive pressure ventilation for two minutes, without adequate response, so orotracheal intubation, invasive mechanical ventilation (IMV) and transfer to the Neonatal Intensive Care Unit (NICU) were performed.

On the eighth day, the respiratory pattern worsened, and due to risk factors and prolonged oxygen, management with broad-spectrum antibiotics was initiated.

On the tenth day, he persisted with respiratory distress syndrome (RDS) dependent on IMV, without improvement, and in view of the chest X-ray that persisted with diaphragmatic elevation, a diaphragmatic ultrasound was requested with evidence of right hemidiaphragmatic elevation (Image 1 and Image 2), with diagnostic impression of diaphragmatic paralysis, so pediatric surgery was requested, who considered that a surgical procedure was required, after completion of DART scheme and weight gain.

Image 1. Right diaphragmatic elevation on chest radiography.

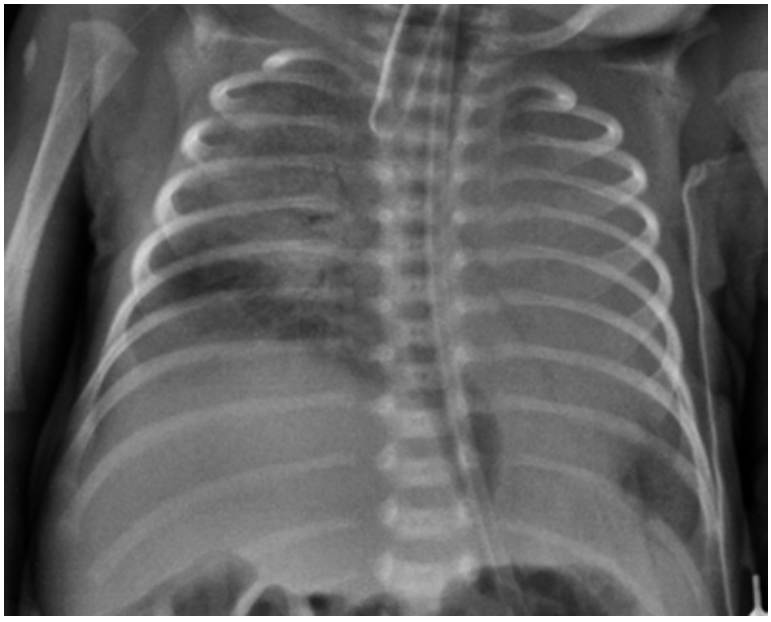
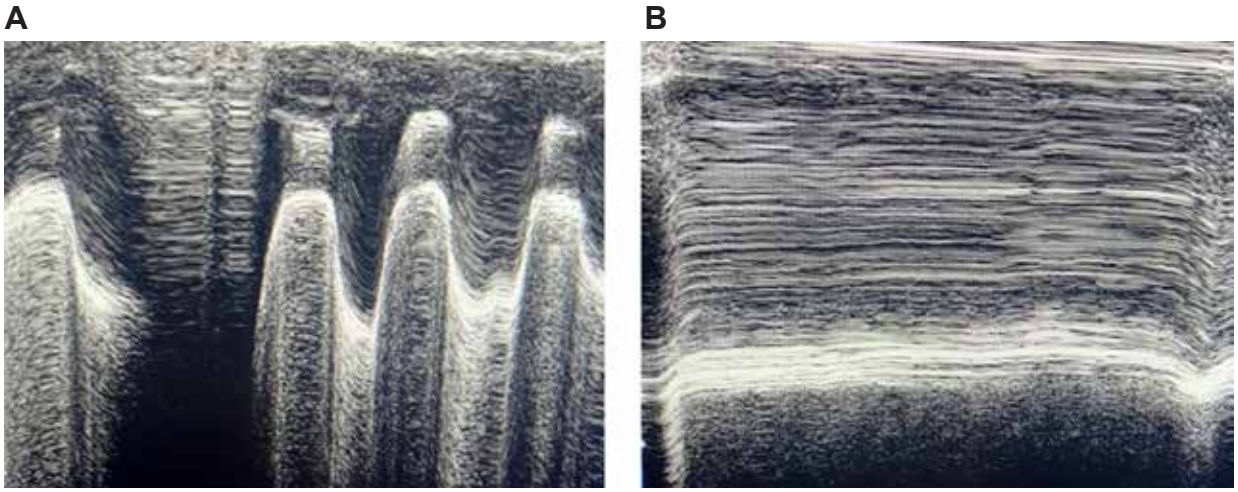
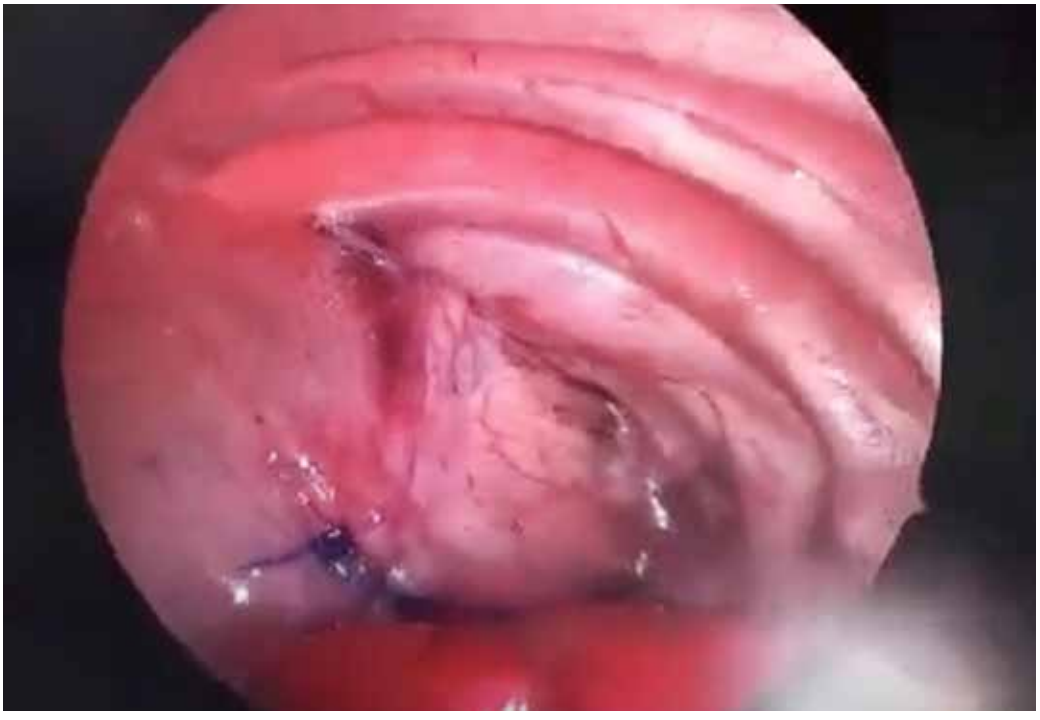


Image 2. Usual movement of left diaphragm (A) and absence of mobility of right diaphragm (B).



On day 25, the evolution was torpid, and despite the low weight, pediatric surgery decided to perform an emergency surgical procedure with intraoperative finding of right diaphragmatic eventration with generalized flaccidity, which rose to the fourth intercostal space, performed anteroposterior plication with three separate stitches (Image 3), descending to the eighth intercostal space, ending the procedure without complications.

Image 3. Diaphragmatic plication is seen, with suture to the diaphragmatic wall towards the eighth intercostal space and two sutures to the diaphragm.



At three months and 17 days of age, he was discharged from the hospital with home oxygen, which could be withdrawn at five months of age.

Discussion

Diaphragmatic eventration is characterized by abnormal elevation of a portion or all of the hemidiaphragm due to lack of muscle or nerve function. This results in weakness and thinning of the affected diaphragmatic portion, leading to reduced function. Depending on the severity, patients may be asymptomatic or present with severe respiratory distress (6).

It is important to make an early diagnosis of this pathology due to possible life-threatening complications. This diagnosis is made by means of radiography, another confirmatory diagnostic method is ultrasound; treatment in mild cases is usually with supportive care, when hypoxemia is present, sometimes oxygen by nasal cannula is not enough, because continuous pressure in the airway (CPAP) is required. The most severe cases require mechanical ventilation and steroids; diaphragmatic plication is reserved for cases that do not respond to this last strategy, that is, when it is not possible to withdraw the patient from mechanical ventilation or when there are other indications (2), see table 1.

Table 1. Indications for surgical intervention.

Indications for surgery: diaphragmatic plication of diaphragmatic plication
Respiratory distress that does not respond to conservative treatment
Dyspnea that is not due to another process (CHF or primary lung disease)
Infants with inadequate nutritional intake or growth retardation
Recurrent or life-threatening pneumonia
Inability to be weaned from mechanical ventilation.

The purpose of diaphragmatic plication is to minimize the abundant diaphragmatic surface and lower the hemidiaphragm (5). In our patient, the weight secondary to prematurity made surgical intervention necessary; in newborns with adequate birth weight, surgery is the last strategy to be used and is elective. Additionally, it is important to highlight that despite the weight of the newborn, it was possible to perform a minimally invasive approach by thoracoscopy with adequate anesthesia tolerance and without immediate or late complications.

Conclusions

Congenital diaphragmatic eventration involves challenges that demand a rapid and effective response. A multidisciplinary approach is essential to ensure the best possible outcome. In addition, the application of minimally invasive surgical techniques is crucial to perform precise and less invasive procedures. In this case, surgery emerges as an essential measure, especially given the patient's prematurity and severe respiratory complications. It is important to emphasize the skill of the surgical team in advanced minimally invasive surgical techniques. Finally, it is of great relevance to keep in mind that this type of patient requires comprehensive and coordinated care to optimize their treatment.

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