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BRIEF REPORT

Congenital lobar hyperinflation: Conservative management as an alternative therapy☆

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KEYWORDS
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Abstract Congenital lobar emphysema used to be treated surgically. Congenital lobar hyperinflation is the currently recommended term, as it involves pathologically healthy lung tissue, which is why conservative management may be an option. Four cases of diagnosed congenital lobar hyperinflation are presented in which conservative treatment was chosen due to their clinical stability. Their outcome has been satisfactory with progressively normal radiology.

PALABRAS CLAVE
Malformaciones broncopulmonares;
Enfisema;
Tratamiento

Hiperinsuflación lobar congénita: manejo conservador como alternativa terapéutica

Resumen El enfisema lobar congénito suele tratarse quirúrgicamente. Actualmente, se recomienda el término de hiperinsuflación lobar congénita, ya que se trata de un tejido pulmonar anatomopatológicamente sano, motivo por el que el manejo conservador puede ser una alternativa válida. Se presentan 4 casos diagnosticados de hiperinsuflación lobar congénita en los que se optó por el tratamiento conservador debido a su estabilidad clínica y en los que la evolución de los mismos ha sido satisfactoria con normalidad radiológica progresiva.

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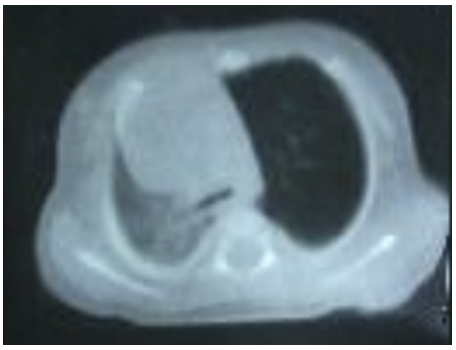

Figure 1 Chest CT scan: hyperinflation of the LUL with mediastinal shift and collapse of contralateral lung.

Introduction

Congenital malformations of the lung and airways constitute a broad spectrum of developmental anomalies, some of which may remain asymptomatic and be discovered accidentally in imaging studies.[1]

Although they are not rare, their frequency is difficult to determine precisely; percentages from 7.5 to 18.7% have been published.[2] They are responsible for a substantial amount of morbidity in neonates, infants, children and even adults and represent the second leading cause of early mortality among infants after anomalies of the cardiovascular system.[3]

Congenital lobar emphysema or hyperinflation is a malformation related to a cartilage or connective tissue anomaly (50%). This disorder gives rise to a ball-valve mechanism permitting inflow of air but obstructing outflow.[2] It arises most frequently in the upper lobe of either lung and occasionally in the middle lobe of the right lung or in other areas, and can be observed from birth.[4]

Surgery (lobectomy) is usually considered the first-choice treatment in symptomatic patients.[1]

After reviewing clinical histories (with prior authorisation from the parents and/or guardians), we present four cases which were managed conservatively, not surgically, given the patients' clinical stability.

Clinical cases

Patient 1

An infant aged 3 months, with no clinical history of interest, attended the accident and emergency department with signs of respiratory distress, preceded by catarrh of the upper air passages on the previous days. The chest X-ray and CT scan showed an area of lobar hyperinflation at the left upper lobe (LUL) (Fig. 1).

Once the manifestations of acute respiratory infection had been resolved, she became asymptomatic, and it was decided to pursue conservative treatment with follow-up at a paediatric respiratory clinic.

Her evolution was satisfactory, with no further acute clinical exacerbations. Chest radiology was normal at age 6. She remains asymptomatic at 17, with normal respiratory function tests.

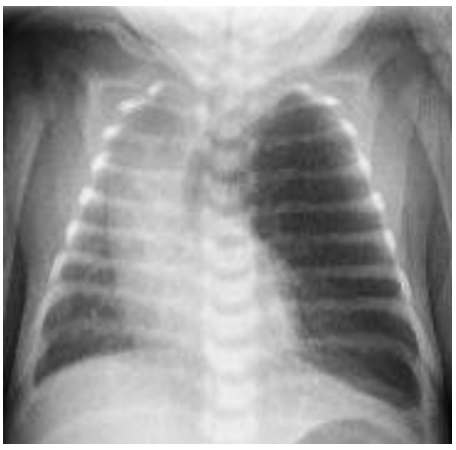

Figure 2 Chest X-ray: hyperinflation of the LUL with contralateral mediastinal shift.

Patient 2

Newborn male aged 20 days, with no personal history of interest, attended the accident and emergency department after 24 h of presenting symptoms characterised by weakness, vomiting, fever, and test results compatible with urinary tract infection. No respiratory symptoms reported apart from noisy breathing. Clinically stable, with adequate tolerance to oral intake and no respiratory compromise, and hypoventilation observed in upper left field. A chest X-ray showed hyperinflation of the left hemithorax with contralateral mediastinal shift (Fig. 2); the CT scan confirmed a marked LUL hyperinflation with constriction of vessels compatible with obstructive emphysema, indicative of lobar emphysema of the LUL.

The study was completed by performing flexible bronchoscopy, revealing a left main bronchus that was structurally and functionally normal, except at the distal level (carina at bifurcation of LUL and lingula), where a collapse of the bronchial lumen – estimated at 80–90% – was observed, due to redundant mucosa and substantial malacia. Given the absence of respiratory effects on the patient, after resolving the infectious process which motivated his admission it was decided to discharge him from hospital with conservative treatment of the emphysema and follow-up of his clinical evolution. The patient is currently 7 months old, is asymptomatic and attends the paediatric respiratory clinic for periodic checkups.

Patient 3

Infant aged 3 months, with no personal or family history of interest, attended the accident and emergency department after 5 days of showing symptoms consisting of frequent congested cough, fever of up to 38.8 °C in the last 48 h, increasing respiratory distress and refusal of food in the last few hours. In the examination, the most notable finding was a tachypnoea of 60 bpm with sub- and intercostal retractions and with wheezing and SaO_2 to ambient air of 92%; respiratory auscultation revealed global hypoventilation with crackling in both pulmonary fields. The chest X-ray showed widespread hyperinflation in both lungs and LUL,