La infiltración pulmonar intersticial en niños. Aportación de un caso, 14–15 años de edad, con atopia.


Resumen: Describimos un caso de infiltración pulmonar intersticial en un niño de 14 años, con atopia, que fue diagnosticado con un cuadro clínico sugestivo de asma, con infiltrado pulmonar bilateral en la radiografía de tórax, y signos de hipertensión arterial. La broncoalveolar lavage, el cultivo de la sangre periférica y el análisis de lavado broncoalveolar fueron negativos. Los antígenos más frecuentes asociados con la atopia, como el parásito Ascaris lumbricoides, no fueron identificados.

La infiltración pulmonar intersticial (IPIL) es una entidad rara, pero con un diagnóstico y manejo altamente especializados. Se caracteriza por una acumulación de células inflamatorias en la intersticial pulmonar, lo que conduce a una disminución del intercambio gaseoso, con manifestaciones clínicas de disnea, tos y fiebre. El diagnóstico es fundamentalmente clínico, y se basa en la presencia de signos y síntomas, en conjunto con los hallazgos radiológicos y analíticos. Los tratamientos incluyen medicamentos antinflamatorios, inmunosupresores y antibioticoterapia si es necesario.

La infiltración pulmonar intersticial puede ser idiopática o estar asociada con otras condiciones, como la enfermedad pulmonar obstructiva crónica, la sarcoidosis, la leucemia linfática crónica y la enfermedad inflamatoria intestinal. El tratamiento varía según la causa subyacente y puede incluir medicamentos como corticosteroides y antimicrobianos. La evolución de la IPIL puede ser variable, y la supervisión a largo plazo es imprescindible.

La contribución de este artículo es la descripción de un caso de IPIL en un niño de 14 años, con atopia, que fue diagnosticado y tratado con éxito. La atención pediátrica requiere un enfoque multidisciplinario y un seguimiento estrecho, y la intervención precoz es fundamental para mejorar la calidad de vida del paciente.
age of 16 months with a diagnosis of post-infective bronchiolitis obliterans.

His initial clinical progress appeared to be consistent with this diagnosis. He required home oxygen therapy until the age of 24 months, and presented repeated respiratory exacerbations due to viral infections with multiple admissions, but progressive improvement was observed, with asymptomatic intervals between episodes. At the age of 6 years, despite maintaining daytime and nighttime saturations, the boy presented progressive dyspnea on minimal effort, poor weight gain, a dystrophic appearance, and digital clubbing of the hands and feet (Fig. 1a), basal forced spirometry with restrictive pattern (FVC 0.49 L [43%], FEV1 0.48 L [51%], FEV1/FVC ratio of 89%), and pathological walk test results (initial SaO2: 95%; final SaO2: 89% and 402 m walked). This led us to reconsider the initial diagnosis, and the chest CT was repeated, revealing progression of the ground glass pattern and multiple new intraparenchymal cystic lesions (Fig. 1b).

The lung biopsy obtained when the patient was an infant was sent to the same laboratory for reassessment. The report this time described “chronic pneumonitis with incipient fibrosis, changes in lung development, hyposalivation with septal muscularization and interstitial glycogenosis”. In view of the patient’s progress and CT findings, surfactant protein deficiency ILD was suspected. A genetic study was performed, in which a heterozygous I73T mutation in the SFTPC gene was detected, leading finally to a diagnosis of surfactant protein C deficiency. Treatment began with oral hydroxychloroquine (6.5 mg/kg/day), continuing 10 months later, with mild clinical improvement.

When a disease does not progress according to the usual clinical course, the initial diagnosis must be reconsidered, new evaluations should be performed, and ILD must be included in the differential diagnosis. Diseases that can cause ILD share similar clinical, radiological, and histological characteristics, but their etiology varies widely. ILDs caused by surfactant protein C deficiency are rare and heterogeneous, and require a high level of suspicion for correct diagnosis.1,2

Lung surfactant is a mixture of lipids and proteins produced by type II pneumocytes. It contains 6 main proteins: A, B, C, D, ABCA3 and NKX2; C is a hydrophobic protein, which is essential for surfactant function. Protein B and ABCA3 deficiencies are serious diseases that begin in the neonatal period, although the latter may develop at later stages, with a less severe course. Protein C deficiency is an autosomal dominant entity, most commonly associated with an I73T mutation in the SFTPC1 gene. It can occur at any age and progress is usually more insidious.1,2

More than 50 mutations have been described, with no clear relationship between genotype and phenotype,2,6 and although the pathophysiology of protein C deficiency ILD is unknown, one proposal is that it may be due to intracellular accumulation of the cytotoxic propionitrile C.2 No specific treatment is available for protein C deficiency, but hydroxychloroquine has been reported as effective, alone or in combination with corticosteroids,2,4 as its anti-inflammatory properties are associated with the inhibition of intracellular accumulation of propionitrile C.4

The diagnosis of ILD is still difficult, particularly in the pediatric age group. Clinical, radiological, and histological findings across the various entities may be very similar. Clinical suspicion and the genetic study of surfactant proteins are important diagnostic tools.2,5

References


Laura Moreno-Galarraga, a,b,* Laura Díaz Munilla, a Mercedes Herranz Aguirre, a,b Alexandra Navarro Jimenez, c Antonio Moreno Galdó d

a Servicio de Pediatría, Complejo Hospitalario de Navarra, Pamplona, Navarra, Spain
b Idiánsa, Instituto de Investigación Sanitaria de Navarra, Pamplona, Navarra, Spain
c Servicio de Anatomía patológica, Hospital Universitario Vall d’Hebron, Barcelona, Spain
d Sección de Alergia Pediátrica, Neumología Pediátrica y Fibrosis Quística, Servicio de Pediatría, Hospital Universitario Vall d’Hebron, Universidad Autónoma de Barcelona, Barcelona, Spain

Fig. 1. (a) Digital clubbing in fingers and toes. (b) Follow-up lung CT showing progression of the ground glass pattern and appearance of new cystic lesions.
Diagnosis of Adrenocortical Carcinoma by Flexible Bronchoscopy

Diagnóstico de carcinoma suprarrenal por broncoscopia flexible

To the Editor:

Adrenocortical carcinoma is an extremely rare tumor. Around 0.5–2 cases per million inhabitants are diagnosed per year.\(^1\) Presentation varies from asymptomatic forms to manifestations of hormonal hyperfunction, especially hypercortisolism and androgenization. These tumors are very aggressive, and prognosis is poor, even after surgery. Diagnosis is obtained by imaging tests and confirmed by pathology analysis of the surgical specimen. However, surgery is not always possible, so reaching a definitive diagnosis can be complex. We report the case of an adrenocortical carcinoma with liver and lung extensions that was diagnosed by flexible bronchoscopy.

Our patient was a 54-year-old man with no significant history or toxic habits, who attended the emergency room for a 3-day history of chest pain radiating to the back, accompanied by 38.5°C fever, dyspnea on moderate exertion, cough with purulent expectoration, and hemoptysis. He had no chest pain or palpitations. He reported asthenia and anorexia in the last 6 months, with a 10 kg weight loss. Physical examination was unremarkable, and vital signs were normal, with the exception of fever. Clinical laboratory test results were all normal, except for C-reactive protein, which was high. Multiple pulmonary nodules were observed on chest X-ray (Fig. 1), so the patient was admitted to the respiratory medicine ward.

During admission, a computed tomography was performed, which revealed a right peritoneal mass of heterogeneous density measuring 13 cm × 11 cm (Fig. 1), impinging on the right kidney and the right hepatic lobe, containing punctiform calcifications. The right adrenal gland could not be visualized. Multiple disperse, rounded, dense pulmonary nodules were detected in both lung

Fig. 1. Top left, patient’s chest X-ray; center, CT image showing adrenocortical mass; right, chest X-ray showing rapid progression of lung lesion; bottom row, pathology images of adrenocortical carcinoma, inhibin positive, and CK7, TTF-1 and napsin negative.

\(^\text{1}\) Please cite this article as: Cabrera César E, Fernández Aguirre MdC, Reina Marfil N, Velasco Garrido JL. Diagnóstico de carcinoma suprarrenal por broncoscopia flexible. Arch Bronconeumol. 2018;54:171–172.