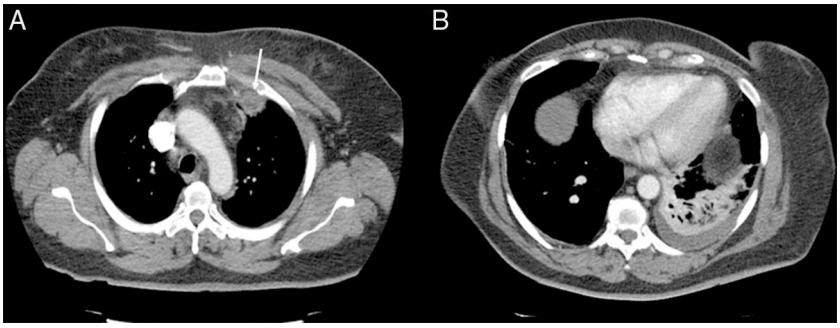

Fig. 1. Pseudonodular image in the space between the first and second left costal arch (arrow), and simultaneous consolidation in the left lower lobe.

costal arch, and pulmonary consolidation with pleural effusion in the left lower lobe (Fig. 1). Findings were confirmed on both ultrasonography of the neck and bone scintigraphy. Ultrasound-guided fine needle aspiration and biopsy was performed, from which *S. aureus* was isolated. The strain was resistant to ampicillin, and susceptible to erythromycin, gentamicin, clindamycin, ciprofloxacin, levofloxacin, and cotrimoxazole. The same microorganism was isolated from the bronchoscopy samples. During admission, intravenous ciprofloxacin and amoxicillin–clavulanic acid were administered, in line with susceptibility results, and improvement was observed in clinical symptoms, radiological signs, and acute phase reactants. Drainage was not required. Treatment continued on an outpatient basis for another 40 days, with complete resolution of the syndrome.

SSA is exceptional and accounts for only 1%–9%[2,4] of SA, and generally occurs in patients with debilitating risk factors and immunosuppression.[1–6] It is also unusual to see the simultaneous development of SA in the acute period of an episode of pneumonia, as it tends to occur later.[1,2] In our patient, the SSA was attributed to the bacteremic pneumonia, as the same microorganism was isolated. *S. aureus* pneumonia in a patient without risk factors is in itself exceptional. The clinical picture of SSA, in contrast to our case, is generally insidious, and presents with fever, pain in the shoulder, and edema and erythema in the sternoclavicular joint.[1,2,4–6] The most widely used diagnostic test is ultrasound, although CT can identify the degree of bone destruction, and scintigraphy is used to delimit the inflammatory area and guide the biopsy and aspiration procedure. The definitive diagnosis depends on isolation of the microorganism. This will indicate the appropriate antibiotic therapy, which should continue for at least 4 weeks in the absence of complications.[1–3,5,6] Surgical treatment is recommended in case of extensive osteomyelitis, abscesses, empyema, or mediastinitis.[1,4,5] In conclusion, pneumonia can unusually cause SA, and exceptionally SAA, and these entities may go unnoticed in the clinical context. As this process is potentially disabling and possibly fatal, etiologic diagnosis should not be delayed.

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1579-2129/

Pediatric Interstitial Lung Disease: An Ongoing Challenge☆

La enfermedad pulmonar intersticial en el niño. Todavía hoy un reto diagnóstico

To the Editor,

Surfactant protein C deficiency causes interstitial lung disease (ILD) of varying severity. Diagnosis in children is complex, due to the rarity and heterogeneous clinical manifestations of this entity.[1,2] We report a case of this disease that was initially incorrectly diagnosed.

Our patient was a boy, born at term, with no significant history. At the age of 14–15 months, he was admitted twice due to bronchiolitis caused by syncytial respiratory virus and bronchitis due to adenovirus. After the second admission, persistent tachypnea, respiratory failure, and bilateral infiltrates on chest X-ray were observed. Further examinations ruled out malformations, immunodeficiencies, pulmonary hypertension, and other infections. Bronchoalveolar lavage was performed: cell count was normal, with no eosinophilia, and negative Gram stain and microbiological cultures. Lung computed tomography (CT) showed a diffuse ground glass pattern with hilar lymphadenopathies. Lung biopsy was performed by thoracoscopy. The specimen was sent to a reference laboratory, and the report described "changes associated with bronchiolitis obliterans". The patient was discharged at the

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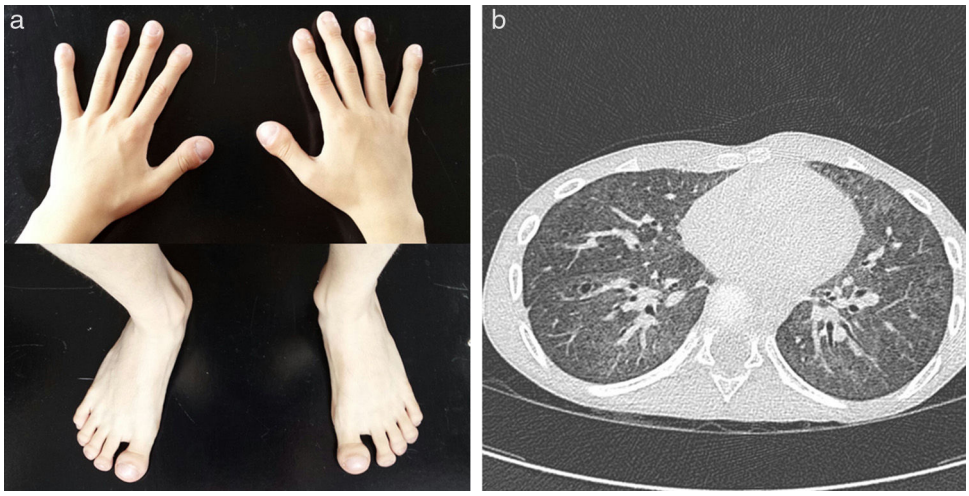

Fig. 1. (a) Digital clubbing in fingers and toes. (b) Follow-up lung CT showing progression of the ground glass pattern and appearance of new cystic lesions.

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%], FEVI 0.48 [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, hypoalveolization 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.[3] 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] 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 proprotein 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 proprotein 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]

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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

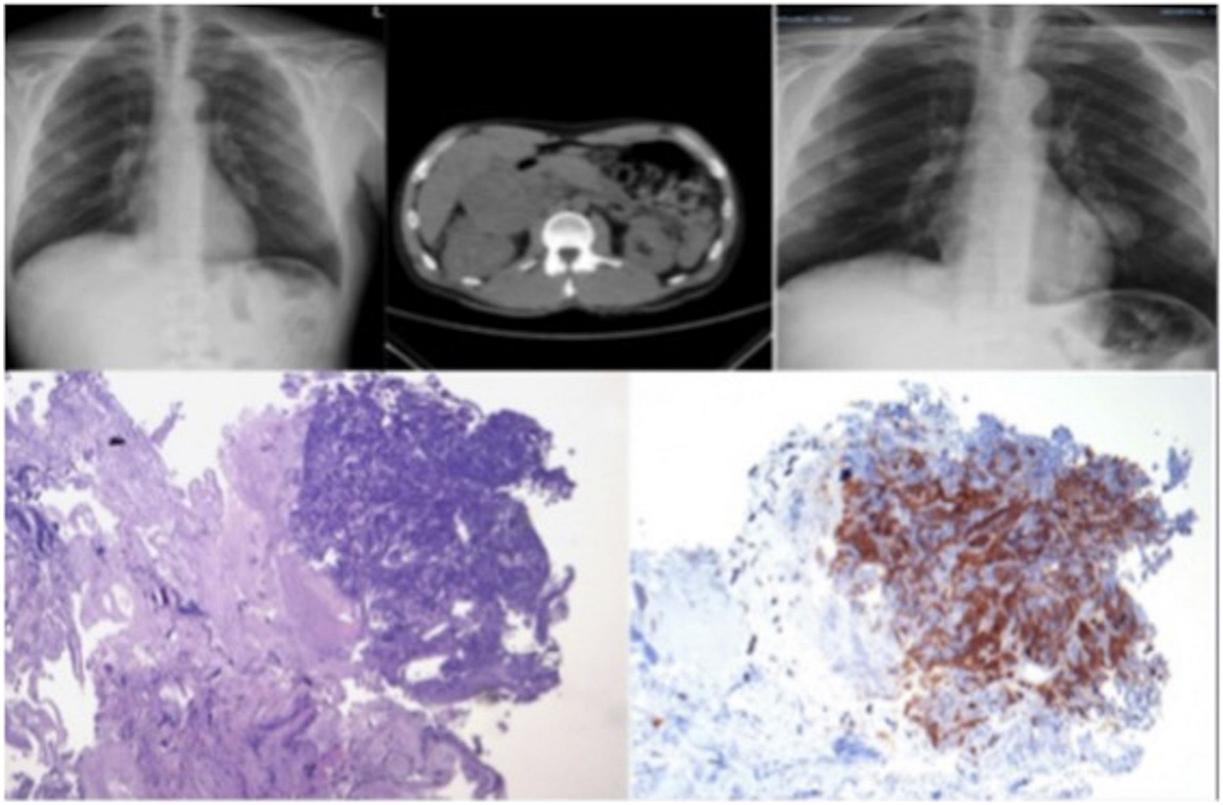

Fig. 1. Top left, patient's chest X-ray; center, CT image showing adrenocortical mass; right, chest X-ray showing rapid progression of lung lesion; bottom row, pathology images of adrenocortical carcinoma, inhibin positive, and CK7, TTF-1 and napsin negative.

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