

Non-invasive Ventilation in Amyotrophic Lateral Sclerosis in Outpatients and Inpatients. Different Perspectives?*



Ventilación no invasiva en la esclerosis lateral amiotrófica en pacientes ambulatorios y hospitalizados, ¿una perspectiva similar o diferente?

To the Editor:

Amyotrophic lateral sclerosis is a neuromuscular disease characterized by progressive motor neuron degeneration. The poor prognosis of this disease means that integrated patient management, including early introduction of non-invasive mechanical ventilation, is essential for improving survival and quality of life.¹

We read with interest the original article by Sanjuán-López et al.,² discussing the importance of integrated pulmonary care and the detection of possible shortcomings. In our opinion, however, some points mentioned in the study need to be addressed:

1. Exclusion of outpatients from the study limits the extrapolation of clinical course and survival outcomes, since by including only patients who needed hospitalization, the authors have selected a subgroup of patients who are probably in a more serious clinical situation than those treated at home. Accordingly, it would have been interesting to include a subgroup of subjects treated on an outpatient basis, specifying the criteria used for initiating non-invasive mechanical ventilation (NIMV) and the progress of these patients.
2. According to the authors, NIMV was initiated in the patients in this study on the basis of respiratory failure criteria, without taking into account the current recommendation for indicating NIMV in patients with ALS on the basis of clinical criteria and lung function tests.³ Indeed, only 30 of the 43 patients who started NIMV during admission had previous lung function test results, suggesting that in many of the cases, NIMV was

introduced at a late stage, with possible repercussions on the clinical course and survival of patients described in the article. This issue is of importance since there are two advantages to initiating NIMV on the basis of an early pulmonary assessment: on the one hand, patients have a better chance of being managed in outpatient clinics, with close collaboration between the home and the hospital⁴; and on the other, it improves quality of life and survival. Thus, hospital admission would be limited to more complex situations, such as NIMV maladjustment or complications derived from disease progression.

In our opinion, the role of the pulmonologists and the involvement of a multi-disciplinary team are essential for reaching an integrated evaluation and an early pulmonary assessment. This in turn optimizes patient management and improves both quality of life and survival.⁵

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Superior Vena Cava Syndrome Caused by an Idiopathic Localized Form of Mediastinal Fibrosis*



Síndrome de vena cava superior secundario a una forma focal idiopática de fibrosis mediastínica

To the Editor:

Mediastinal fibrosis (MF) is one of the most common non-malignant causes of superior vena cava syndrome (SVCS), along with thrombi forming on intravascular devices (central lines, pacemaker cables, etc.). However, the most common cause of SVCS remains malignant disease, particularly lung cancer. In endemic regions, MF is a relatively common complication of infections caused by *Histoplasma capsulatum*, although it is more common in our setting to find idiopathic forms, forms associated with immunoglobulin (Ig) G4 deposition (in the context of IgG4-related diseases,

such as retroperitoneal fibrosis or Riedel's fibrosing thyroiditis), or forms caused by other granulomatous diseases, such as tuberculosis.^{1–4}

We report the case of a 44-year-old man, non-smoker, who presented in our hospital with a 1-month history of progressive clinical symptoms of headache and edema of the face and upper limbs. Chest X-ray showed slight widening of the right paratracheal stripe (Fig. 1A), but no parenchymal opacities or pleural effusion. On physical examination, dilation of the neck veins and edema of the face and cervical spine were the only observations of note. Chest computed tomography (CT) confirmed a soft tissue mass that surrounded the circumference of the upper SVC causing severe stenosis of the lumen (Fig. 1B). No radiological signs of previous tuberculosis infection and no infiltration of other mediastinal or lung structures were observed. Tuberculin testing was negative and the patient had never visited any country in America. IgG4 serum levels were normal. A diagnosis of a localized form of MF was proposed, and confirmed with mediastinoscopy. Histology study revealed a fibrous tissue with chronic inflammatory cell aggregation and abundant collagen bundles, with no signs of vasculitis or granulomas. Biopsy culture was negative for fungi and mycobacteria. The patient was initially treated with systemic corticosteroids and anticoagulants, with progressive clinical improvement. A repeat follow-up CT at 4 weeks (Fig. 1C) showed radiological improvement of the SVC stenosis, so after consultation with the Departments of Thoracic Surgery and Vascular Radiology, need for stent placement or bypass was ruled out.

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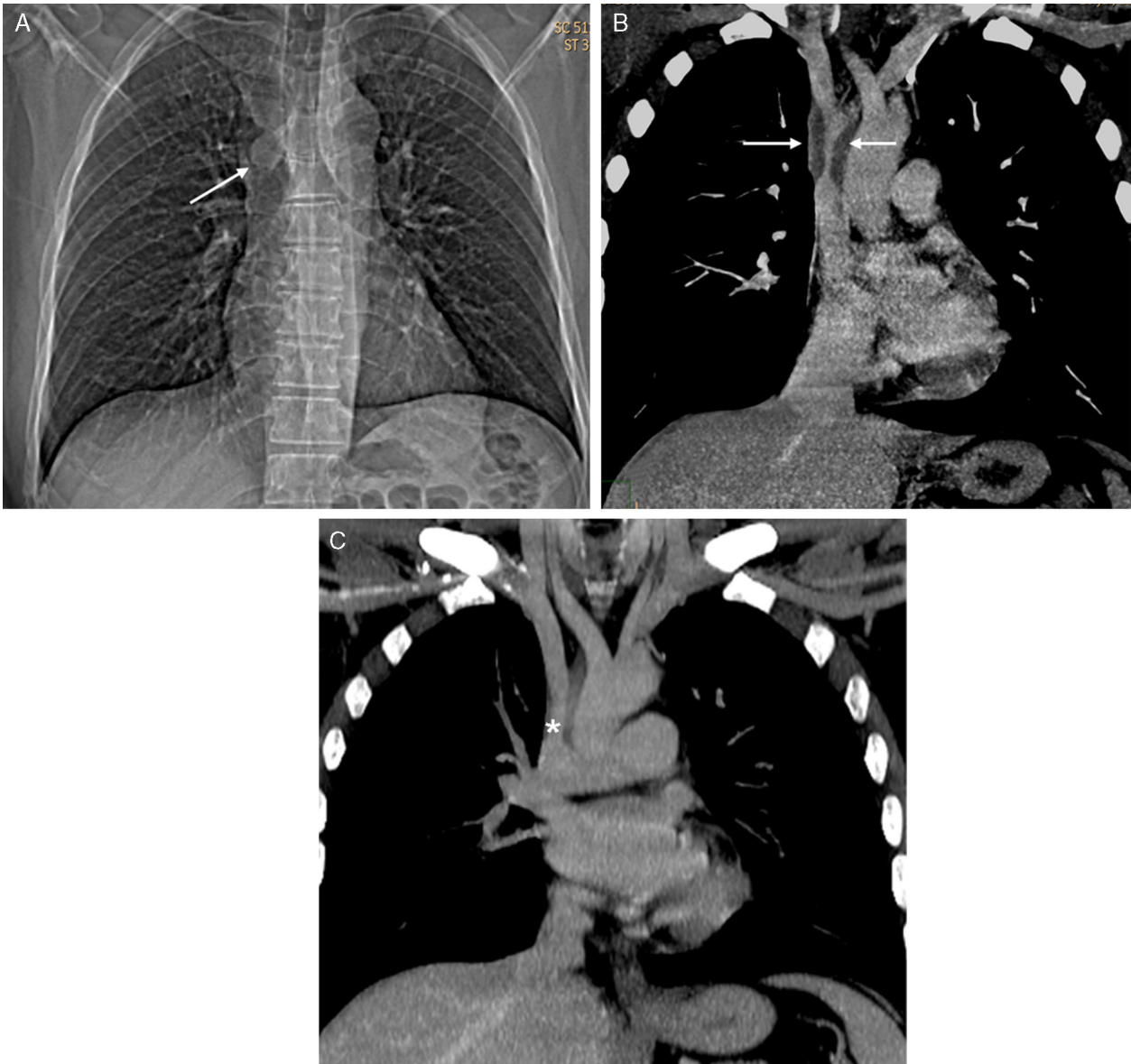


Fig. 1. (A) Chest X-ray showing subtle widening of the paratracheal stripe (arrow). Neither parenchymal opacities nor pleural effusion is observed. (B) Coronal maximum intensity projection (MIP) reconstruction confirming a soft tissue mass surrounding the circumference of the SVC, causing severe stenosis of its lumen (arrows). Coronal MIP reconstruction carried out 4 weeks after starting treatment, showing improvement in SVC stenosis (asterisk).

In our opinion, this case is interesting because it illustrates the importance of imaging studies in the diagnosis, staging and follow-up of an SVCS caused by an idiopathic localized form of MF exclusively affecting the SVC.

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Spontaneous Pneumothorax and Cocaine Use[☆]



Neumotórax espontáneo y consumo de cocaína

To the Editor:

Spontaneous pneumothorax (SP) associated with marijuana or cocaine use is uncommon but not unknown. Although it can be difficult to demonstrate a direct effect, lung damage caused by drug use can predispose patients to pneumothorax.¹ We report the case of a 39-year-old man, referred to our unit for treatment of right SP. He had already had SP in the same side 7 months previously and had admitted to occasional use of cocaine. Mechanical pleurodesis was performed via thoracoscopy with resection of the apex of the right lung. Pathology laboratory analysis showed unexpected evidence of non-necrotizing granulomas in the bronchial walls, associated with small vesicles (Fig. 1). The patient had no significant clinical history and all standard clinical laboratory test results, including mycobacteria, fungal infection and human immunodeficiency virus, were negative.

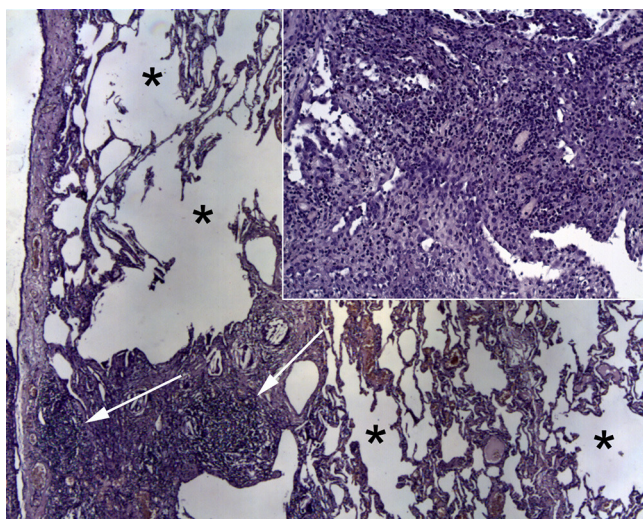


Fig. 1. Pathology sample with hematoxylin and eosin staining (10× magnification) showing granulomas (white arrow) and small vesicles (*). Activated lymphocytes and giant inflammatory cells were observed.

Respiratory tract deposits of particles of talc contained in cocaine were thought to have led to the formation of granulomas as a reaction to a foreign substance. Granuloma growth affected the small airways, causing air retention and bullous disease. Severe cough and bronchospasm caused by the inhalation of cocaine caused increased intra-alveolar pressure, followed by vesicle rupture and pneumothorax.

Ward et al.² and Pare et al.³ described significant radiological changes after cocaine use, including bullous emphysema and pulmonary fibrosis. Our group recently reported vesicles similar to those seen in elderly patients in a series of 13 young habitual marijuana smokers.^{4,5}

Granulomas as the only expression of lung damage may be explained by the fact that our patient reported intermittent and not continuous use of cocaine. However, sustained exposure to cocaine exacerbated the deposit of talc particles, causing severe lung damage, evidenced by the above-mentioned radiological changes.

Granulomas caused by sporadic use of cocaine may predispose the patient to SP, even in the absence of significant radiological changes. The best way of prevent severe parenchymal damage is to avoid the use of drugs.

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