

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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