



Fig. 1. (A) Pulmonary mass in the peripheral right upper lobe (RUL) of necrotic and abscessed appearance, containing a small air bubble (mediastinal window CT image). (B) Previously described nodule has disappeared, and a new cavitated nodule has appeared close by. Bronchial wall thickening and RUL bronchial impaction (lung window CT image). (C) Resolution of previous findings (follow-up lung window CT image).

were detected on CT, described elsewhere as very unusual.⁵ The most common histopathological changes are airway inflammation and bronchiectasis.² Our patient's lung biopsy revealed foci of organizing pneumonia and necrobiotic nodules. These necrotic areas, seen on CT as abscesses, appeared sporadically during our patient's clinical course. To date, flare-ups of organizing pneumonia associated with UC have been described,⁴ but necrobiotic nodules are rarer.² Treatment of UC lung involvement is based on corticosteroids.² Our patient's response to corticosteroids was excellent, with complete clinical and radiological resolution after 2 weeks of treatment.

To conclude, this is a case of pulmonary involvement of UC, with an interesting presentation, due to exacerbations occurring with hemoptysis, respiratory failure and radiological images of migratory cavitated pulmonary nodules, with no associated clinical symptoms. Definitive diagnosis was based on lung biopsy results and response to steroid treatment was complete.

Conflict of Interests

The authors state that they have no direct or indirect conflict of interests with the contents of this manuscript.

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Silvia Barril,^a Ana Rodrigo-Troyano,^a Ana Giménez,^b Oriol Sibila^{a,*}

^a Servei de Pneumologia, Departament de Medicina, Hospital Santa Creu i Sant Pau, Universitat Autònoma de Barcelona, Barcelona, Spain

^b Servei de Radiologia, Departament de Medicina, Hospital de la Santa Creu i Sant Pau, Universitat Autònoma de Barcelona, Barcelona, Spain

* Corresponding author.

E-mail address: osibila@santpau.cat (O. Sibila).

Mortality in Obesity-Hypoventilation Syndrome and Prognostic Risk Factors[☆]



Mortalidad en el síndrome de obesidad-hipoventilación y factores de riesgo pronóstico

To the Editor,

We read with interest the letter sent to ARCHIVOS DE BRONCONEUMOLOGÍA regarding our article "Noninvasive Mechanical Ventilation in Patients With Obesity Hypoventilation Syndrome. Long-term Outcome and Prognostic Factors".¹ We would like to thank the authors and venture to respond.

We agree that the lack of comorbidity data is the greatest limitation of this study, particularly since the main aim was to define prognostic factors for predicting mortality. However, this aspect was not taken into account in the preliminary design of the database, and we rejected the idea of a retrospective search in the clinical records that would have reduced the quality of our

data. While including comorbidities in the analysis would have been interesting, this omission does not affect the results, namely, that patients with sleep apnea and those with better ventilatory function at the start of the ventilation program have the best prognosis.

With regard to the methodological concerns expressed by the authors of the letter, both initiation of ventilation and monitoring of ventilation mode comply with standard recommendations. Lowest pressure support (PS) was 10 cm H₂O, gradually increasing to 16, depending on arterial blood gases and tolerance. If 90% saturation could not be achieved with the initial PS, oxygen supplements were added until saturation was 90%, while FiO₂ was subsequently modified according to arterial blood gas and saturation achieved with the effective or maximum PS. The statistical tests for comparison were selected on the basis of the sample size and normal distribution, and nonparametric tests were used, assuming penalties.

Although all patients were included in the analysis of lung function and gas exchange outcomes until they left the ventilation program, the survival analysis was performed exclusively on patients who remained on ventilation until death (endpoint event). This means that patients who were withdrawn from the program due to poor compliance were not included in these analyses. As the authors of the letter rightly observe, and as confirmed in a

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recently study,² compliance is key to the success of noninvasive ventilation.

In our opinion, nighttime monitoring of patients is particularly important. While we agree that studies evaluating the benefits of polygraphy and/or polysomnography monitoring are needed, we are convinced that ventilator efficacy must be monitored in terms of PaCO₂, the value directly related with alveolar ventilation. Our belief, corroborated by other authors,³ is that this type of monitoring is essential. It is a routine practice in our clinic, and early morning arterial blood gases are measured in the patient on ventilation both at the start of the ventilation program and in all follow-up visits.

In brief, although the study is limited by the lack of data evaluating the impact of comorbidity, we believe that it is important to have been able to show in a long term study that the severity of ventilatory impairment is a factor for poor prognosis in patients with obesity-hypoventilation syndrome requiring noninvasive ventilation, while concomitant sleep apnea constitutes a protective factor. PaCO₂ monitoring is essential for ensuring effective ventilatory support and obtaining good outcomes.

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E. Ojeda Castillejo,* P. de Lucas Ramos, P. Resano Barrios, S. López Martín, P. Rodríguez Rodríguez, L. Morán Caicedo, J.M. Bellón Cano, J.M. Rodríguez González-Moro

Hospital General Universitario Gregorio Marañón, Madrid, Spain

* Corresponding author.

E-mail address: e.ojeda.castillejo@gmail.com (E. Ojeda Castillejo).

Combination of Tracheal Bronchus and Partial Anomalous Pulmonary Venous Return in a Patient With Type 1 Neurofibromatosis and Ipsilateral Vagal Nerve Neurofibroma[☆]



Asociación de bronquio traqueal y drenaje venoso pulmonar anómalo parcial en paciente con neurofibromatosis tipo 1 y neurofibroma del nervio vago ipsilateral

To the Editor:

Type 1 neurofibromatosis (NF1) is the most common form of neurofibromatosis, a group of genetic disorders of the nervous system that mainly affect the development and growth of neural cell tissues. NF1 occurs in approximately 1 of every 3000 newborns. Transmission to progeny is by the autosomal dominant mode, although up to 50% of new cases occur spontaneously through *de novo* mutations. The literature describing the association of NF1 with cardiovascular or airway malformations is sparse.¹

We report a clinical case of a 45-year-old woman with a diagnosis of NF1, non-smoker, with dysphagia due to a posterior mediastinal tumor causing compression of the thoracic esophagus. On magnetic resonance imaging, the tumor was radiologically consistent with right vagal nerve neurogenic tumor. The physical examination only revealed café-au-lait spots on the skin and several scars on her back and abdominal wall from previously resected neurofibromas at these sites. In view of symptoms of progressive dysphagia, we decided to perform surgical resection of the posterior mediastinal tumor. The pre-operative chest X-ray revealed suspected tracheal bronchus (TB) (Fig. 1A), so these examinations were complemented with a chest computed tomography (CT). This

confirmed TB, and revealed the unexpected combination of partial anomalous pulmonary venous return (PAPVR), consisting of drainage from the right upper lobe vein into the superior vena cava (Fig. 1B–D).

The combination of NF1 with pulmonary development anomalies, either vascular or airway, and their diagnosis in adults is exceptional, and, to our knowledge, this the first report of simultaneous NF1, PAPVR and TB.¹ TB is one of the most common abnormalities of the bronchial tree and, despite its name, more often occurs in the right main bronchus than in the trachea itself. TB is generally diagnosed incidentally in asymptomatic patients undergoing bronchoscopy or chest CT for other reasons. It can, however, occasionally cause hemoptysis, recurrent lung infections, or right upper lobe atelectasis in intubated patients in whom the distal end of the endotracheal tube is located at or distal to the origin of the TB.² PAPVR is a vascular development anomaly consisting of abnormal drainage of one or more pulmonary veins into a systemic vein (the innominate vein, superior vena cava, azygos vein, right atrium, coronary sinus, or inferior vena cava), causing left-to-right shunt. Adults with PAPVR have a risk of developing long-term precapillary pulmonary hypertension due to volume overload in the right ventricle.³ The combination of these 2 development anomalies in a single patient is exceptional.⁴ The association of NF1 with pulmonary development anomalies is very rare (2.3%).¹ In a review of vascular and cardiac malformations in 2322 patients with NF1, pulmonary stenosis and aortic coarctation were more prevalent, but no cases of anomalous pulmonary vein drainage (partial or total) were identified.¹ Detection of these 2 anatomical variants may have clinical, anesthetic and therapeutic implications in patients scheduled for thoracic surgery. In our case, the anesthesiologist and the thoracic surgeon were aware of the anomalies, and complete thorascopic resection of the right mediastinal tumor with preservation of the ipsilateral vagal nerve could be planned and successfully performed. Histological examination confirmed a myxoid neurofibroma without atypia. The post-operative period was incident-free, and the patient was discharged 4 days after the intervention.

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