effusion without parenchymal or mediastinal abnormalities. Two percutaneous Abrams needle biopsies of the pleura (with 3 weeks of interval) were negative for malignancy and infection, only demonstrating nonspecific fibrous thickening of the pleura. The thoracoscopy showed hyperaemia and thickening of the costal pleura, and small whitish nodular lesions, that were biopsied (Fig. 1). An apple-green birefringence was noted on Congo-red staining under polarized light, compatible with pleural amyloidosis, mostly with a perivascular pattern (Fig. 2).

Eight months after the thoracoscopy, the pleural effusion recurred. This time, it was bilateral with a moderate volume and a transudate. Despite initiating treatment for WD few months after the beginning of symptoms, the inflammatory process evolved to systemic amyloidosis, which was the major factor contributing to the progressive deterioration of our patient, with cardiac and renal failure. The patient died due to multiple organ failure, after 3 years of follow-up.

Although a definitive diagnosis of WD could not be made, the endoscopic characteristic findings and the response to the antibiotic therapy suggest it as the precursor of the AA amyloidosis.

Pulmonary amyloidosis may present as tracheobronchial infiltration, parenchymal nodules, persistent pleural effusions, and pulmonary hypertension.[3,4] Pleural involvement is very rarely reported, and it is usually associated with AL amyloidosis, which accounts for up to 80% of pulmonary amyloidosis.[5,6] Typical aspects of thoracoscopy consist of hyperaemia of the pleural surface, inflammation with nodular lesions or brown nodules of the parietal pleura.[5,7] Persistent pleural effusions occur in 1–2% of patients with systemic amyloidosis and are usually associated with poor prognosis and often refractory to treatment. Pleurodesis has been effective in some cases.[7,8]

This case illustrates the difficulty in diagnosing pleural amyloidosis, after two failed needle pleural biopsies. This is probably due to the fact that amyloid deposition is not uniform over the pleural surface. The pleural effusion in our patient may derive from increased fluid production induced by inflammation, interruption of lymphatic drainage caused by amyloid infiltration of the pleura, decreased resorption of fluid from the pleural space due to vascular deposition of amyloid and in the bilateral pleural effusion from congestive heart failure secondary to amyloid infiltration in the heart.[5,7]

Although in this case the previous diagnosis of amyloidosis supported and incited to actively search for amyloidosis, it should be always kept in mind, as pleural effusion is a rare manifestation of pulmonary involvement but could be the presenting manifestation of amyloidosis.

Conflicts of interest

The authors have no conflicts of interest to declare.

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Use of the Intermittent Abdominal Pressure Ventilation to guarantee speech in a tracheostomized Amyotrophic Lateral Sclerosis patient

Dear Editor,

Amyotrophic Lateral Sclerosis (ALS) is a fatal, progressive, neurodegenerative disease. When Respiratory failure is too severe to be corrected with Non Invasive Ventilation and/or when bronchial secretions cannot be managed with noninvasive techniques, tracheostomy and invasive mechanical ventilation (IV) are an option.[1]

Tracheostomy ventilation significantly prolongs survival in ALS patients without effect on the disease progression. For this reason, patients with IV experience a worsening of disability and an increment of the dependency with a severe impairment of their quality of life.[2] One of the most important aspect of the multidisciplinary approach in ALS is to guarantee as long as possible the maintenance of the residual functions. In this context, the first and most widely used strategy to allow tracheostomized patients without severe bulbar involvement to speak is the simple cuff deflation, but, in a percentage of these patients, this technique fails

Table 1 Ventilation and cough machine settings (PS: pressure support; IP: inspiration pressure; EP: expiration pressure; RR: respiratory rate; Ti: inpiratory time; Te: expiratory time; Tp: pause time; Tr Insp: inspiratory trigger; IV: invasive ventilation; TOV: tracheal open ventilation; IAPV: intermittent abdominal pressure ventilation; APCV: assisted pressure-control ventilation; ST: spontaneous timed).

	IV	TOV	IAPV	Cough machine
Mode	APCV	ST	APCV	Auto
PS (cmH_2O)	13	16	40	
IP				40
Peep (cmH_2O)	4	4	0	
EP				40
RR	14	14	18	
Ti (s)	1.4	1.4	2.2 (Te)	3
Te (s)				2
Tp (s)				1
Tr Insp	High	High	Auto	
Rise time (ms)	400	400	600	

with the consequent impossibility of verbal communication and a severe reactive mood depression.[3]

Intermittent Abdominal Pressure Ventilation consists of an elastic inflatable bladder incorporated within a corset surrounding the abdomen. With bladder inflation by a ventilator, the abdominal content and diaphragm move upward, assisting expiration. With bladder deflation, inspiration occurs passively. There have been only scattered reports on the use of IAPV[4–7] and two publications concerning its use in large populations of patients, in a regimen of noninvasive[8] and invasive[9] ventilator support.

A 49 years old man with a diagnosis of definite ALS, according to El Escorial Criteria, was admitted for the first time to our Centre, having recently had tracheostomy and gastrostomy done because of worsening in respiratory function and swallowing, The patient complained of difficulty using home IV device and difficult management of the secretions. NIV was introduced in Spring 2014 after onset of wheezing during hospitalization in an intensive care ward of an other Centre. The patient reported intermittent use of non invasive ventilation due to poor tolerance until Autumn 2014. At that time there was a significant worsening of respiratory involvement due to a right lung pneumonia associated to weight loss and deterioration of swallowing and dysphagia.

During the hospitalization, we optimized both IV and secretion clearance by cough machine through the tracheostomy tube in association with tracheal aspirations. He could not sustain the spontaneous breathing, so, to permit speech, we introduced diurnal tracheal open ventilation (TOV) with cuff deflation and the concomitant usage of a speaking valve, but with poor patient tolerance due to discomfort with the unnatural experience of air coming in the upper airways and poor synchrony with the ventilation. The patient could not even manage secretions and saliva. Thus, to increase the feasibility and pleasure of speaking during mechanical ventilation and improve breathing comfort we introduced diurnal IAPV (*Pneobelt*™) associated with a tracheostomy speaking valve for spontaneous breathing. During tracheal ventilation, our patient was fitted while supine,

Figure 1 Pneobelt™ corset.

with the corset's horizontal upper border approximately two finger breadths below the costophrenic junction. Once positioned, the patient was placed in a wheelchair, his cannula wad deflated and a speaking valve was placed. *Pneobelt*™ was connected to the same portable ventilator used for tracheal ventilation but set with parameters ''*ad hoc*''for IAPV.

Table 1 shows IV, cough machine and IAPV settings (Fig. 1) shows Pneobelt™ with the corset and inner bladder.

Table 2 Pulmonary gas exchange (ABG: arterial blood gas analysis; IV: invasive ventilation; IAPV: Intermittent Abdominal Pressure Ventilation).

ABG first evaluation	IV	IAPV + speaking valve
PH	7.57	7.43
PaO_2	84.2	71
$PaCO_2$	36.3	40
HCO_3^-	27	27