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Pleural effusion in AA amyloidosis - A rare involvement of a rare disease

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in each stage of its manufacture.<sup>2</sup> Wood smoke is intensely released from the kilns, and represents a complex mixture of liquid, solid and gaseous particles, irritant and genotoxic, such as nitrogen and sulphur oxides, benzene, methanol, styrene, phenols, naphthalene, aldehydes, organic acids and polycyclic aromatic hydrocarbons.<sup>2</sup>

Both superior and inferior airway symptoms have been reported, including sneezing, nasal secretion, cough, expectoration, dyspnea, wheezing and even hemoptysis related to lower airway disease.<sup>3</sup> Rhinitis, asthma, chronic obstructive pulmonary disease and respiratory infections may be observed in such workers.<sup>3</sup> Literature is scant concerning interstitial lung disease/pneumoconiosis related to wood charcoal dust, especially regarding the HRCT findings. De Capitani et al.<sup>4</sup> reported 3 cases of pneumoconiosis related to wood charcoal dust and activated carbon dust exposure. In all cases, HRCT descriptions match those observed in our cases, with centrilobular ground glass nodules. Our finding of air trapping on the expiratory scan probably representing small airway disease, was not mentioned in De Capitani et al.<sup>4</sup> report. Similar findings have been described in carbon black pneumoconiosis, related to burning of natural gas and various petroleum products, with diffuse centrilobular ground glass nodules.<sup>5</sup> We found no other papers describing HRCT appearance on charcoal dust exposure.

Histopathological description of black pigmented macules was observed both in our patient as in two of De Capitani et al.<sup>4</sup> patients. No significant findings of fibrosis or inflammatory disease were observed in our patient's transbronchial or surgical biopsy, supporting the non-fibrogenic nature of this pneumoconiosis.<sup>4</sup>

In conclusion, we describe clinical, tomographic and histopathologic features of this uncommon interstitial lung pneumoconiosis related to wood charcoal smoke exposure. Considering the fact that biomass fuels are broadly disseminated as energy sources, especially in developing countries, prevention strategies in charcoal manufacturing should be encouraged in order to prevent such complications.

## Conflicts of interest

The authors have no conflicts of interest to declare.

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## Pleural effusion in AA amyloidosis – A rare involvement of a rare disease

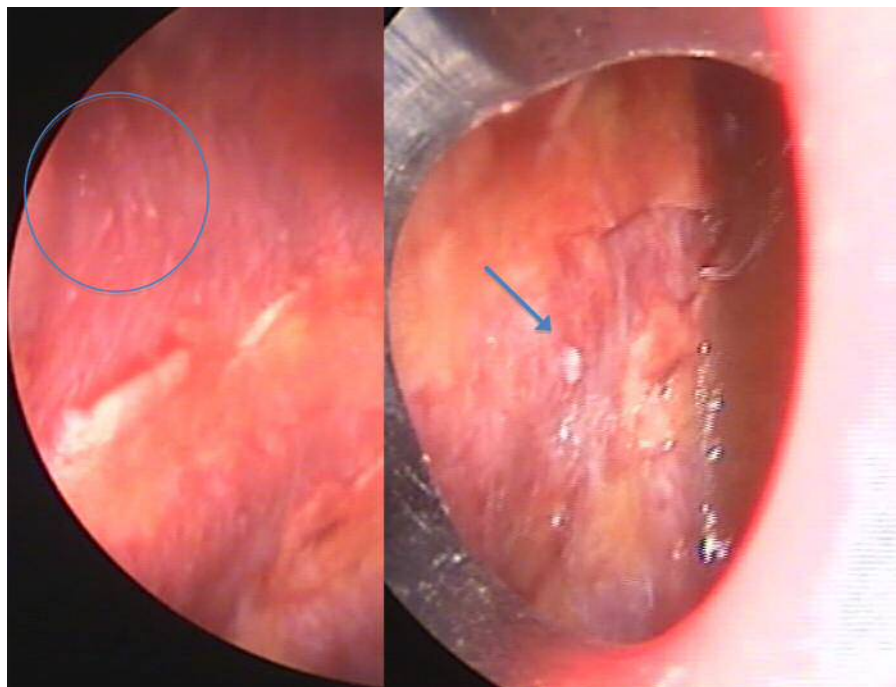


Dear Editor,

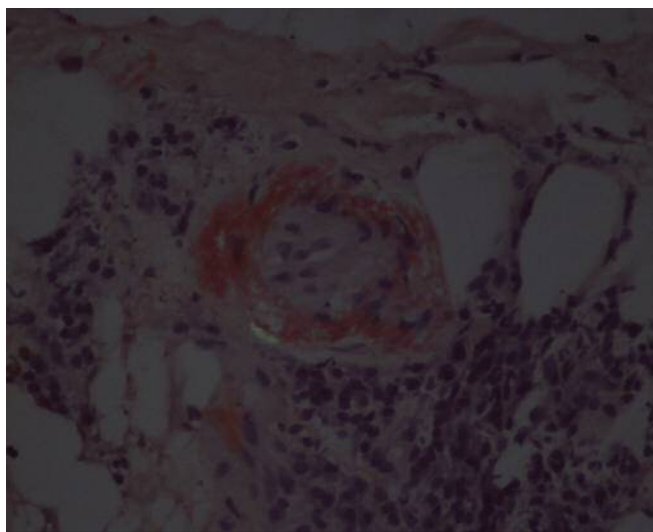
Amyloidosis are a group of heterogeneous diseases due to protein misfolding and amyloid deposition in various organs and tissues, resulting in a wide range of clinical manifestations depending upon their chemical composition, location, and amount. The most frequent types of amyloidosis are the immunoglobulin light chain (AL) amyloidosis and the serum amyloid A, an acute phase reactant (AA) amyloidosis. The former is associated with plasma cell dyscrasia, while the latter results from longstanding chronic inflammatory/infectious diseases.<sup>1</sup> Reports about amyloid involvement of the respiratory tract are scarce.

We report the case of a 52-year-old female with type 2 diabetes mellitus and arterial hypertension who was admitted due to a 6-month history of anorexia, non-bloody diarrhea and weight loss. Upper gastrointestinal endoscopy revealed numerous lymphangiectasias in duodenum mucosa. Histological staining of duodenum biopsy samples showed macrophages containing bacilliform structures positive for periodic acid-Schiff (PAS) staining. The classic diagnosis of Whipple disease (WD) is based on positive PAS staining of duodenal biopsies, but PAS staining can be positive in other circumstances, such as *Mycobacterium* infection.<sup>2</sup> Real time PCR assays for *M. avium* and *M. intracellulare* were negative, and fungal structures were not identified in the biopsy specimen.

Despite the negative PCR for *Tropheryma whippelii*, the suggestive histological findings (but not entirely diagnostic) and the endoscopic characteristic observation, WD was assumed and the patient was treated accordingly. She



**Figure 1** Thoracoscopy reveals small whitish nodular lesions at the costal pleura (circle, arrow).



**Figure 2** Parietal pleura at polarized light, there is an apple green refringence, confirming the deposition of amyloid substance (magnification  $\times 400$ ).

completed 15 days of intravenous ceftriaxone 2 g/od followed by oral cotrimoxazole (CTX) 960 mg/bid with clinical improvement. One month later, she presented with acute kidney disease – creatinine 3.54 mg/dL [normal range (NR), 0.5–0.9 mg/dL], urea 180 mg/dL (NR, 10–50 mg/dL), hypoalbuminemia 17.5 g/L (NR, 38–51 g/L) proteinuria 194 g/24 h (NR, <0.1 g/24 h) and CTX was changed to doxycycline 100 mg bid and hydroxychloroquine 200 mg bid was initiated. A kidney biopsy revealed serum amyloid A (AA) amyloidosis. Due to effort dyspnea, an echocardiogram was performed and showed a normal cardiac function (left ventricular ejection fraction 67%) and signs suggestive of infiltrative cardiomyopathy. During a 2-year follow-up, there

was a stabilization of renal function with conservative measures (creatinine 3–3.5 mg/dL) and no recurrence of gastrointestinal symptoms. Surveillance endoscopies disclosed persistence of lesions suggestive of WD.

After this period, a decrease in breath sounds was noted over the right lung base. The patient was asymptomatic, afebrile, without peripheral edema. A chest radiograph revealed a mild right pleural effusion. A diagnostic thoracentesis showed a clear light yellow fluid, exudate, glucose 118 mg/dL and normal ADA. Cytological analysis showed a mononuclear predominance. Pleural fluid study was negative for malignancy or infection (*T. whipplei* PCR was negative). Thoracic CT scan revealed unilateral pleural

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.<sup>3,4</sup> Pleural involvement is very rarely reported, and it is usually associated with AL amyloidosis, which accounts for up to 80% of pulmonary amyloidosis.<sup>5,6</sup> Typical aspects of thoracoscopy consist of hyperaemia of the pleural surface, inflammation with nodular lesions or brown nodules of the parietal pleura.<sup>5,7</sup> 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.<sup>7,8</sup>

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.<sup>5,7</sup>

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.<sup>1</sup>

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.<sup>2</sup> 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