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Figure 1. Chest high-resolution CT, coronal slice: diffuse reticulo nodular infiltrate, with calcifications in some lesions.

thoracoscopy, showed the presence of nodular foci of osseous metaplasia.

Two weeks after the biopsy, the patient developed edema of the lower limbs below the knee. Urine testing showed proteinuria 3.6 g/24 h and hypoalbuminemia 2.8 g. Nephrotic syndrome was diagnosed and a renal biopsy was performed, consistent with light-chain (AL) amyloidosis, with lambda light chain as a precursor of amyloid. Lambda chains of 723 mg/L (5.7–26) were found in blood. A bone marrow biopsy showed 40% plasma cells, confirming the diagnosis of multiple myeloma.

In view of these findings, the lung biopsy was reviewed and found to be positive for red Congo and birefringence both in the blood vessel walls and the alveolar septa, confirming lung involvement in the form of pulmonary alveolar septal amyloidosis.

Treatment started with bortezomib and glucocorticoids. An echocardiogram was repeated 2 months after the diagnosis, showing an increase in refringence in the myocardium and endocardium of both ventricles, consistent with infiltrative cardiomyopathy. The patient died 1 month later due to cardiac decompensation.

Pulmonary alveolar septal amyloidosis is characterized by the deposition of amyloid in the alveolar septa and blood vessels. In general, it is a component of systemic AL amyloidosis compromise, and is most often caused by the deposit of lambda light chains. Several case series report that pulmonary alveolar septal involvement occurs in 78–90% of cases of AL amyloidosis.2,3

The clinical presentation of pulmonary alveolar septal amyloidosis is more severe than with other types of pulmonary amyloidosis, as the deposits occur in the interstitium and affect gas exchange. It presents as progressive dyspnea, not attributable to other causes. This is reflected in lung function tests that show a restrictive pattern with a reduced diffusion capacity for CO and hypoxemia on exertion.4

High-resolution CT shows reticulo nodular opacities, septal thickening, and less frequently, ground glass opacification, bronchiectasis, and honeycombing.5

Histologically, amyloid may adopt an interstitial nodular pattern that can diffuse or form plaques. It can also be seen in the vascular walls. Interstitial deposits can go unnoticed. As in this case, calcifications and foci of osseous metaplasia can be observed.6

The treatment of this disease is the same as for systemic AL amyloidosis. Reducing light chain concentrations in blood may improve organ dysfunction.7 However, no data are available on the impact of treatment on pulmonary compromise.

References


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