Incidental multiple adenopathies and the diagnosis of systemic amyloidosis

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CASE REPORT

This report describes the clinical case of a 67-year-old man with hypertension referenced to the medical consultation for the study of several bilateral mediastinal adenopathies discovered accidentally on a chest computed tomography (CT) following the patient’s admission to the emergency department after falling off a tree. Of note in the physical examination is the presence of macroglossia and thick lips (Figure 1A).

In the complementary study of adenopathies serologic data and a positron emission tomography (PET) scan were requested. Analytically with a sedimentation rate of 21 mm, the peripheral blood smear analysis was normal and there was a good renal function with no change in phosphocalcic metabolism. At the level of immunoglobulins, a value of IGG-2300 mg/dl was found with the respective gamma-monoclonal peak (IGG-lambda) in the electrophoresis allowing the diagnosis of monoclonal gammopathy of undetermined significance.

Posteriorly, PET scan demonstrated additional cervical and mediastinum-hilar bilateral ganglia with low metabolic activity of inflammatory nature, without significant hypermetabolism of 18F-FDG (Figure 1B, C). The definitive diagnosis of primary amyloidosis was obtained later through abdominal fat biopsy (Figure 1D). An echocardiogram was requested to rule out the cardiac involvement of the disease. Given the clinical stability of the patient and the absence of significant findings in the complementary diagnostic tests, the strategy adopted in this patient was “watch and see.”

DISCUSSION

Amyloidosis is a heterogeneous group of disorders characterized by extracellular deposition of abnormal protein in a β-pleated configuration that makes it resistant to proteolysis. It can be classified according to its biochemical profile as either primary or secondary. Primary amyloidosis occurs less frequently than the reactive or immune-related secondary form. Regarding the anatomical location, amyloidosis can be classified as localized disease (10–20%) or systemic disease (80–90%) [1].

Lymph node involvement occurs in up to 37% of patients with systemic amyloidosis and the hilar, mediastinal, and para-aortic lymph nodes are most commonly involved, in contrast localized primary amyloidosis presenting as lymphadenopathy is a very rare entity.

Respiratory involvement is observed in about half of patients with amyloidosis, and cases with the highest clinical significance are seen in patients with primary amyloidosis [2].

Regarding lung disease patterns, they are divided between the standard lymphatic, diffuse alveolar septal, nodular, and tracheobronchial parenchyma. The lymphatic and septal patterns are most commonly seen in patients with systemic involvement, and nodular and tracheobronchial parenchymal patterns are more commonly seen in localized disease [3, 4].

CONCLUSION

Thoracic lymphadenopathy, alone or with interstitial disease, is the most common CT finding in primary...
amyloidosis with systemic involvement, not described in secondary form. On the other hand, pulmonary involvement with hilar and mediastinal lymph node enlargement is uncommon in localized amyloidosis.

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**Keywords:** Adenopathies, Monoclonal gammopathy of undetermined significance, Systemic amyloidosis

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João Costelha – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Artur Barros – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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