A Case of Pulmonary Amyloidosis with Progressive Systemic Involvement

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A Case of Pulmonary Amyloidosis with Progressive Systemic Involvement

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Introduction

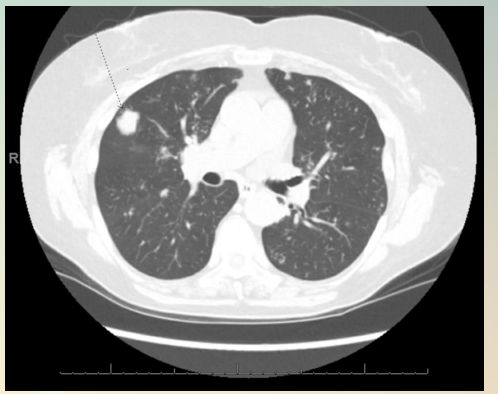
- Amyloidosis is a group of disorders characterized by abnormal deposition of fibrillar proteins into beta-pleated sheet configuration.
- Pulmonary manifestation of amyloidosis may be localized to the lungs or may be a part of the systemic disease.

Case

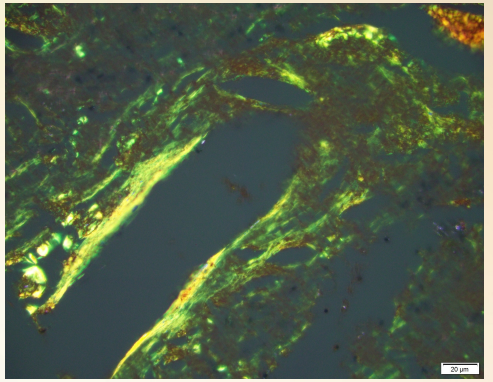
- 79-year-old female with a past medical history of Sjögren Syndrome, discoid lupus and Ramsay-Hunt syndrome was found to have numerous bilateral pulmonary nodules as incidental findings on chest x-ray and computed tomography (CT) of the chest.
- She underwent wedge resections and tissue pathology with Congo red stain, which demonstrated apple-green birefringence of the eosinophilic material under polarized microscopy.
- Serum electrophoresis revealed a spike in the gamma region, consistent with IgG monoclonal gammopathy.
- A year later, CT chest revealed increased size of bilateral pulmonary nodules. Repeat CT five months later noted enlarging mediastinal lymph node.
- Patient subsequently underwent transbronchial needle aspiration, which was negative for malignancy.
- Repeat CT imaging again noted increased size of bilateral pulmonary nodules and patient underwent right lung core needle biopsy to differentiate between AL amyloidosis and AA amyloidosis.
- Biopsy confirmed pulmonary nodular amyloidosis with mass spectrometry further classifying as AL amyloidosis.
- Due to rising NT-proBNP suggesting cardiac involvement, patient was started on bortezomib-based chemotherapy for systemic amyloidosis approximately 26 months into diagnosis.
- Patient has been responding to treatment with reduction in free light chain and NT-proBNP. Additionally, she is undergoing evaluation at Hershey Penn State for autologous stem cell transplant.

Discussion

- Diagnosis of amyloidosis is challenging as amyloid proteins can infiltrate virtually any organ system and have diverse clinical presentation.
- Patients who are asymptomatic with localized amyloidosis do not require chemotherapy.
- Awareness of systemic amyloidosis is essential as these patients benefit from autologous stem cell transplant and chemotherapy.



High Resolution CT demonstrates numerous pulmonary nodules. One of the more prominent nodules is located in the right upper lobe measuring 1.9 x1.4 cm.



This image displays apple-green birefringence using Congo red stain under polarized light characteristic of Amyloidosis. Further studies classified it as AL (lambda)-type amyloid deposition.

REFERENCES

Vieira IG, Marchiori E, Zanetti G, et al. Pulmonary amyloidosis with calcified nodules and masses - a six-year computed tomography follow-up: a case report. *Cases J*. 2009;2:6540. Published 2009 Sep 8. doi:10.4076/1757-1626-2-6540

Gandham AK, Gayathri AR, Sundararajan L. Pulmonary amyloidosis: A case series. *Lung India*. 2019;36(3):229-232. doi:10.4103/lungindia.lungindia_205_18

Milani P, Basset M, Russo F, Foli A, Palladini G, Merlini G. The lung in amyloidosis. Eur Respir Rev. 2017;26(145).

Mahmood S, Bridoux F, Venner CP, et al. Natural history and outcomes in localised immunoglobulin light-chain amyloidosis: a long-term observational study. Lancet Haematol. 2015;2(6):e241-250.

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