Pulmonary Nodular Amyloidosis in a Patient with Sjogren Syndrome

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Image in Rheumatology

A 61-year-old woman was admitted for assessment of multiple nodules incidentally detected on chest radiography (Figure 1A). Six months previously, we diagnosed Sjögren’s syndrome upon observation of symptoms of dryness of the mouth and eyes, positive Schirmer’s test, positive fluorescent antinuclear anti-

Figure 1. Multiple nodules and a mass in both lung field. Histology showing dense accumulation of typical amorphous eosinophilic material and calcification, and Congo red stain showed apple-green birefringence.
body test, elevated levels of anti-Ro antibodies, and positive
salivary gland scintigraphy findings. Spirometry showed moder-
ate restrictive ventilatory defect (FVC: 1.9 L, 65% of predicted,
DLco: 70%). Chest tomography revealed a mass measuring
4.7×7.8 cm on the right lower lobe, multiple calcified nodules
in both lungs (Figure 1B). Fluorodeoxyglucose positron emis-
sion tomography showed multiple hypermetabolic amorphous
masses (Figure 1C). Lung tissue specimens showed dense accu-
mulation of typical amorphous eosinophilic material and calcifi-
cation (Figure 1D, hematoxylin-eosin stain, 40×), and Congo
red staining showed apple-green birefringence (Figure 1E).
Immunohistochemistry using specific antibodies to the λ light
chain confirmed the diagnosis of light-chain (AL) (λ) amylolosis. Amyloidosis rarely causes diffuse nodular amyloi-
dosis and exclude associated lymphoma. The treatment of dif-
fuse nodular amyloidosis in SS is complicated by its rarity (2).
We suggested treatment with high-dose steroids followed by or-
al or parenteral cyclophosphamide, but the patient did not
agree. We performed computed tomography (CT) every 6
months for 1 year; during this period, her condition remained
unchanged, without any significant deterioration or evidence of
systemic amyloidosis or lymphoma. Clinicians should be aware
that nodular or cystic lung lesions in primary SS represent amy-
loidosis, and a lung biopsy must be performed for confirmation.

References
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