Clinical History:

A 80 year old man presented with multiple bilateral pulmonary nodules, progressively enlarging and multiplying during the last three years.

Imaging Findings:

A 80 year old man presented with multiple bilateral pulmonary nodules, progressively enlarging and multiplying during the last three years. No other complaints were reported and no other clinical signs were revealed during physical examination. There was no history of drug ingestion and no significal past medical history. Laboratory tests including complete blood count, serum chemistry and arterial blood gases revealed normal results. The patient's serum analysis showed IgG lambda type monoclonal gammopathy. No Bence-Jones protein was found in 24h urine collections. Pulmonary function tests showed slight obstruction with normal diffusion. Bronchoscopy revealed a normal tracheobronchial tree. Bronchoalveolar lavage was negative for malignancy. Transthoracic fine-needle aspiration of pulmonary nodules was negative, twice during these years. Thyroid, heart and upper abdomen ultrasonographic examination was normal. Histological examination of gastric and bowel mucosa was negative for amyloid or active inflammatory disease. Open-lung biopsy and microscopically examination showed that pulmonary nodules were composed of waxy eosinophilic material. The diagnosis of amyloidosis was made histologically by the finding of apple-green birefringence under polarized light, in Congo red stained sections. Staining was abolished by pretreatment of the sections with potassium permanganate. The presence of lambda light chains showed that it was AL in origin.

Discussion:

Amyloidosis is a group of diseases due to deposition of insoluble protein fibrils, or proteins complexes with polysaccharide material in connective tissue, around parenchymal tissue cells and in the walls of blood vessels. The disease can be either localized (10% to 20% of cases) or systemic (80% of cases). Systemic form is classified as primary or secondary. Secondary type is found in patients with chronic inflammatory disease (such as tuberculosis, osteomyelitis, bronchiectasis, rheumatoid arthritis, leprosies, Crohn's disease, ankylosing spondylitis, Reiter's syndrome, psoriatic arthritis, chronic rheumatic heart disease, dermatomyositis, scleroderma, Behcet's syndrome and rarely in systemic lupus erythematosus), in patients with familiar Mediterranean fever and in those with malignant neoplasm (such as Hodgkin's disease and hypernephroma). In 75% of cases of the generalized disease there are amyloid deposits in the mucosa of rectal biopsy specimens. In the rare primary type of the disease, amyloid is deposited in the hurt, gastrointestinal tract, lungs, muscle and skin. It develops in patients with no other
disease and in those with plasma-cell abnormalities or abnormal immunoglobulins. Amyloid deposition of primary form is of light chain protein type known as AL amyloidosis. Systemic or reactive amyloidosis known as AA amyloidosis (due to protein A amyloid deposition) is the acquired disorder that may develop in patients with chronic infections or inflammatory diseases. AA amyloidosis can be associated with generalized organ involvement such as spleen, liver, adrenal glands and kidneys, with renal involvement producing the earliest manifestations. Amorphous amyloid protein material with the microscopic appearance of paraffin infiltrating between cells, when exposed to Congo red stain and vied under polarized light microscope shows apple-green birefringence. Our report shows the natural radiological 3 years history of an almost clinically asymptomatic primary pulmonary nodular AL amyloidosis, in an 80 years old male patient, with no other organ involvement. Primary nodular pulmonary amyloidosis is an uncommon manifestation. In this form amyloid is deposited in the alveolar walls and around the intralveolar capillaries, as well as in the walls of the smaller blood vessels in the lung. Deposits may also occur in bronchical and tracheal walls. The disease runs a benign course, but usually becomes a diagnostic problem due to nonspecific histological features. Most pulmonary deposits are asymptomatic, some may become manifested as nodular shadows that may be up to several cm in size easily identified on chest roentgenograms or CT scans in a peripheral or sudpleural location. The shadows are growing slowly over the years and may cavitate or become calcified. The nodular form of amyloidosis may show contrast enhancement on CT and MRI. Occasionally amyloid deposition is confined to hilar and mediastinal lymph nodes. Pleural effusion may be present. Radiological differential diagnosis must include metastatic disease, abscesses, granulomatous disease, rheumatoid lung, sarcoidosis, mucoid impaction and fungal disease.

**Differential Diagnosis List:** Primary pulmonary nodular amyloidosis, with no other organ involvement.

**Final Diagnosis:** Primary pulmonary nodular amyloidosis, with no other organ involvement.

**References:**

Ikeda S, Takabayashi Y, Maejima Y, Tachibana N, Ehara T, Nezu A, Hoshii Y.

Mathews V, Vasudevan AR, Nair S, Cherian AM.

Fitzer PM.

Yoshida T, Obara A, Yamauchi K, Nakadate T, Shiba A, Ohura M, Inoue H, Tomichi N.
Figure 1

Description: A solitary small, 2 cm in diameter, nodule is seen above the left hilus. Origin:
Figure 2

Description: Bilateral relatively poorly-defined nodular lesions, up to 2 cm in diameter, are seen in the middle lung fields. Origin:
**Figure 3**

*Description:* At least three soft tissue attenuation nodules are depicted in the right lung, one of which is punctured. Pay attention to two smaller nodules in asubpleural location bilaterally. *Origin:*
**Description:** Increase in number and size of right sided pulmonary nodules. Pay attention to some small new nodules in the right lower lung fields. Subtle hyper aeration of the left lung is also evident. Twenty months after the first x-ray, considerable advance of the disease is evident. **Origin:**
Description: One soft tissue attenuation nodule is depicted in the left lung field. Origin:
**Description:** A large left sided pleural effusion is present. Note the large mass based on the left pleura at aortic arch level. **Origin:**
Description: Aspiration of the large mass, abutting the left pleura surface. Origin:
Description: Large left pleural effusion in addition to bilateral poorly-defined mass lesions. Hyperlucent area of right upper lung is evident. Origin:
Description: Only resolution of the left sided pleural effusion is seen, meanwhile lung and pleural lesions are remaining. Origin: