Three Cases of the Nodular Pulmonary Amyloidosis with A long-term Observation

Hidekazu Suzuki, Kaoru Matsui, Tomonori Hirashima, Masashi Kobayashi, Sinji Sasada, Norio Okamoto, Naoko Kitai, Kunimitsu Kawahara, Haruyuki Fukuda, Takefumi Komiya and Ichiro Kawase

Abstract

Long-term observation with chest radiograph and computed tomography (CT) scan was performed for pulmonary amyloidosis. There are few reports of primary pulmonary amyloidosis with a long-term observation. We encountered three cases of nodular pulmonary amyloidosis observed by intermittent chest radiograph or CT for 5 years or more. The patients were a 54-year-old man, and 67- and 68-year old women. For diagnosis, transbronchial biopsy and percutaneous lung biopsy were performed. Amyloid nodules grew slowly and two cases showed findings of cavity and calcification.

Key words: pulmonary amyloidosis, Long-term observation, Bronchofiberscopy

Introduction

Amyloidosis is defined as an abnormal protein formed by amyloid fibrils that are deposited in the extracellular tissue. Tracheobronchial-pulmonary amyloidosis is cryptogenic and also confined in the respiratory system. Compared with systemic amyloidosis, localized pulmonary amyloidosis usually has a benign course. However, there are few detailed reports regarding the long-term radiographic change. Here, we present three cases of nodular pulmonary amyloidosis that were observed for at least five years.

Case Reports

Case 1

A 68-year-old woman had an abnormal shadow on chest radiograph in a screening survey (Fig. 1). She was a non-smoker and in good health. Her first chest CT scan was performed on February 26, 1999 (Fig. 2a). A single nodule with a calcification of 25 mm in diameter was observed at the right middle lobe. After that, she did not visit the clinic regularly. She came for an examination on September 5, 2003 due to dyspnea on exertion. The chest CT revealed that the nodule was growing (Fig. 2b) and calcification increased. Amyloid deposition was proven by bronchoscopic lung biopsy. Amyloid was not found in the liver or rectum biopsy. The latest CT on March 9, 2005 showed that the nodule size had increased (Fig. 2c). New nodules were observed on the latest CT film.

Case 2

A 67-year-old woman had a check-up on January 23, 1995 chiefly for shortness of breath. She was a non-smoker. She had surgical resection of uterine cancer in 1970. Chest radiograph showed multiple nodules of the bilateral lung (Fig. 3a). They were of clear and round form. Bronchoscopy was incomplete for diagnosis. Percutaneous lung biopsy (needle biopsy) was performed and the sample demonstrated AL type amyloid deposition. The echocardiogram was normal and there was no amyloid in the rectum tissue. All nodules were gradually growing and the numbers had increased. A small cavity was noted in 1999 and had disappeared in...
Figure 1. Chest radiograph findings of a 68-years-old woman (Case 1) on the first visit in 1999. Single nodular shadow size of 22 mm of diameter was noted on the left lower lobe.

Figure 2a, 2b, 2c. Chest CT scan of Case 1. Tumor size was 25 × 15 mm on Feb. 26, 1999 (2a), 25 × 20 mm on Sep. 5, 2003 (2b) and 25 × 23 on Mar. 9, 2005 (2c). The size of the nodule became enlarged from 25 × 15 mm to 25 × 23 mm in six years.

2001 (Figs. 3b, 3c).

Case 3

In a 54-year-old man, an abnormal shadow of chest radiograph was noted in a health examination on August 21, 1991 (Figs. 4, 5a). He had previously been in good health. He worked in the aluminum processing industry for about 20 years. He smoked 1 pack of cigarettes per day for about 34 years. Percutaneous lung biopsy was taken for lesion of the left lower lobe. However, the diagnosis was condensed exudate of the left pleural cavity. Needle biopsy was performed again on June 22, 1992, because the peripheral tumor in the right upper lobe was enlarged. Amyloid deposition was proven in this specimen. No amyloid deposition was observed in the rectum biopsy specimen. Two years later (July 22, 1994), the nodular shadow in the right lower lobe had expanded (Fig. 5b). Two coin lesions newly appeared in the right lung. Bronchoscopic biopsy was performed for the right S10 lesion. This sample documented amyloid deposition. The nodule sizes had increased (Fig. 5c) and the latest CT (Fig. 5d) showed a cavitary lesion and calcification.

Discussion

Nodular amyloidosis is not associated with systemic disease and is associated with a benign prognosis reticulonodular pattern (1) and many cases of pulmonary localized type
are reported to be the AL type of amyloid proteins (2, 3). By observing the deposited locus, it can be divided into two types: localized amyloidosis which deposits only in a specific organ and systemic type with deposition in every organ. Gillmore and Hawkins (3) classified laryngeal, tracheobronchial, parenchymal nodular, diffuse alveolar septal and intrathoracic lymphadenopathy in amyloidosis of the respiratory system. The present three cases were regarded as localized parenchymal nodular amyloidosis and long-term survival was expected. Two of the three cases were asymptomatic and were detected by mass screening. Usually, a patient with pulmonary nodular type amyloidosis has no symptom except for the chest radiograph abnormal shadow and an early symptom is dyspnea (4). It is difficult to suppose that the respiratory symptoms are caused only by peripheral nodules, we presumed that the patient was affected by smoking or other complications. However, we could not prove the affect of smoking and or any complicated disease in our cases. On the other hand, the tracheobronchial type can lead to hemoptysis, bloody phlegm (5) and the intrabronchial polyp can be cauterized by YAG laser (1, 4). Generally, systemic amyloidosis is a very poor prognosis. Utz et al (1) reported that the average survival is 16 months after diagnosis.

There is insufficient data on pulmonary localized amyloidosis that can take a good course with no treatment; in one report, the nodule gradually became enlarged over 14 years, finally occupying all lung fields (6). Slow continual growth of more than 20 years on chest radiograph and asymptomatic cases has been reported (7). As for the characteristic of pulmonary localized amyloidosis, Gillmore and Hawkins (3) claim that nodules are usually peripheral and subpleural, occurring in the lower lobes, which might be bilateral and range in diameter of 0.4-15 cm. In addition, they grow slowly and frequently had cavities or calcification. These reports did not include chest CT but mainly observation by chest radiograph. In the present cases, the most enlarged nodules (Case 3) were observed from 1 mm to 25 mm in diameter at 13 years and cavitary lesion appeared after 4 years (Case 2), and at 13 years (Case 3) after the first visit. In

Figure 4. Chest X-ray of 67-years-old woman (Case 3) on Aug. 21, 1991 revealed multiple bilateral nodules.

Figure 5a, 5b, 5c, 5d. The changes in the chest CT findings of Case 3 shown in 5a (1991), 5b (1994) and 5c (1996). The tumor shadows were larger and increased. The mass of the left lower lobe revealed cavity lesions and calcification (arrows) in 2004 (5d).
case 2, the cavitary lesion disappeared and shrunk in 2001. We supposed that the tumor wall became thinner and was crushed by pressure. On chest radiograph, the findings are somewhat limited. In case 3, there was a lesion like a fungus ball. There was a possibility that this change was modified by fungal infection.

The deposition of amyloid protein in limited areas was due to low pH and neighboring proliferation of monoclonal plasma cells and interaction of glycoaminoglycan (8). Hiroshima et al (9) reported one case in which a significant amount of asbestos was observed around the pulmonary amyloid nodule. They suggest that asbestos inhalation is an etiological factor in amyloidosis.

Bronchoscopic diagnosis was attempted in most cases in Japanese case reports from 1985, but the success rate is low. The common diagnostic means was a surgical technique such as open lung biopsy or resection. There was a successful report for accurate diagnosis with CT-guided bronchoscopy for small lesions (10). In addition, there is the question of serious possible bleeding with amyloidosis. Two of 3 patients in the present paper were proved as amyloidosis by bronchoscopy. Strange et al (11) reported a fatal complication of pulmonary hemorrhage and arterial air embolism after transbronchial biopsy. They proposed that biopsy is a potential risk for amyloidosis. In a Mayo Clinic report, 2 of 11 patients lost approximately 100 mL of blood by bronchoscopic lung biopsy (1). Because amyloid deposition exists in the peri-vascular area with wall thickness or destruction, it may bleed easier than other neoplastic lesions. We think that the examination, when amyloidosis is doubtful, should be cautious. In addition, even if we initially diagnose as amyloidosis, careful follow-up is necessary.

References


© 2006 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imindex.html