Primary Amyloidosis Involving Mediastinal and Hilar Lymph Nodes: A Case Report

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Amyloidosis is a rare systemic disease caused by extracellular accumulation of insoluble fibrillar material in various tissues. The radiographic findings of amyloidosis involving the lymph nodes have not been previously reported in Korea, and we report a rare case of primary amyloidosis involving the mediastinal and hilar lymph nodes, with CT and radiographic appearances.

Index Words: Amyloidosis, Mediastinum, CT, Lymphatic system, diseases

CASE REPORT

A 55-year-old man was admitted to our hospital with symptoms of transient ischemic attack. On routine chest radiograph, incidental findings of soft tissue masses in the AP window, the left hilum and the infradilar region were noted (Fig. 1). Subsequent CT scan of the chest demonstrated multiple enlarged lymph nodes in the left hilum and the mediastinal regions (Fig. 2a, b); some showed tiny punctate calcifications. These nodes were slightly hypodense than the adjacent vascular structures, and were relatively well enhanced after the infusion of contrast medium. The patient had neither symptoms nor past history of any disease. Follow-up chest CT scan nine months later revealed no appreciable interval changes of previously noted lesions. The patient underwent a percutaneous transthoracic CT-guided biopsy, and on hematoxilin-eosin staining, the specimen showed homogeneous eosinophilic materials. Subsequent Congo red staining disclosed amyloid materials with typical apple-green birefringence under polarized light (Fig. 3).

DISCUSSION

The amyloid diseases constitute a group of conditions with diverse causes characterized by the accumulation of ultrastructurally fibrillar material in various tissues. The majority of cases are superimposed on multiple myeloma or other chronic underlying conditions such as tuberculosis, cystic fibrosis, bronchiectasis, lung abscess, rheumatoid disease, syphilis, or Hodgkin's disease. A smaller percentage are primary or heredofamilial in origin. Although they are usually seen in a systemic form, 10–20% of cases can be localized. Males are affected more than females and mean age at presentation is 55 to 60 years (1–4).

The microscopic identification of an amyloid is facilitated by its affinity for Congo red stain. Under polarized light, Congo red imparts to an amyloid a unique apple-green birefringence (2, 5, 6), as in the case of our patient. Because he had no associated underlying chronic disease, we believed that our case was a primary form.

In order to facilitate radiologic discussion, Himmelfarb et al. (7) classified cardiopulmonary amyloidosis...
According to five types: nodular, diffuse parenchymal, tracheobronchial, mediastinal and hilar adenopathy, and cardiac. Pathologically, amyloidosis affects thoracic lymph nodes in almost 20% of cases (3, 4); radiographically, lymph node enlargement is less frequent (3, 5, 8). The radiographic appearance of amyloid involvement of lymph nodes is not well documented; a few reports described the contour of calcifications within the involved lymph nodes: stippled, coarse, diffuse or eggshell-like patterns on radiographs (3, 5, 7, 9).

Fig. 1. A 55-year-old man with amyloidosis involving the mediastinal and hilar lymph nodes. Chest radiograph shows soft tissue masses in the left hilar and infr hilar regions, and the AP window. (AP: aortopulmonary)

Fig. 2. Calcifications within the enlarged mediastinal and hilar lymph nodes.

a. Postcontrast CT scan at the level of the aortic arch demonstrates relatively well enhancing several enlarged lymph nodes in the AP window.

b. Prominently enlarged lymph nodes with multiple punctate calcifications (arrows) are noted in the left hilar region, the azygoesophageal recess, and the retrocardiac portion. Subcarinal lymph node is also enlarged with calcifications (not shown).

Fig. 3. Amyloid materials emitting birefringence. CT-guided biopsy specimen with Congo red staining shows virtual complete replacement of lymph node tissue by amyloid materials with typical apple-green birefringence (arrows) under the polarized light (×100).

and a speckled appearance on CT (8). Although punctate calcifications were also noted in our case, their significance is not certain. The differential diagnosis for these findings includes tuberculosis, sarcoidosis, pneumoconiosis, lymphoma, and rarely, metastasis and Castleman’s disease. Enlarged lymph nodes were relatively well homogeneously enhanced in our case although there was a previous report of inhomogeneous enhancement of the enlarged lymph nodes (10).

The rarity of amyloidosis causes this entity to be omitted from consideration in cases of lymph node enlargement with or without calcification (5, 6). A radiologist should be aware of the possibility of amyloidosis.
in cases with enlarged hilar or mediastinal lymph nodes, particularly in the presence of calcifications.

REFERENCES

제목: Koala Sign

Fig. 1. Large pituitary adenoma on coronal T1WI

Fig. 2. Closed view of the mass

Fig. 3. Koala

설 명: Sellar MRI coronal T1WI에서 large pituitary adenoma가 우연히 코알라와 흡사한 영상을 보게 되었습니다. Mass는 suprasellar growing을 하면서 both carotid artery을 encase하지만 compression하지 않아 코알라의 두눈을 만들고, sphenoid sinus의 일부가 코알라의 코를 형성하고, 위로자란 bulky한 mass가 귀를 만들었습니다. 왼쪽 귀의 일부에는 hemorrhage가 있어 high signal intensity로 관찰됩니다.

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