

Primary Amyloidosis Involving Mediastinal and Hilar Lymph Nodes : A Case Report¹

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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

Amyloidosis is a rare disease complex characterized by diffuse extracellular deposition of an insoluble fibrillar protein material. Radiographic findings of this disease have not been previously documented in Korea, and we report radiographic and computed tomographic(CT) manifestations of the chest in a patient with pathologically proven amyloidosis involving the mediastinal and hilar lymph nodes.

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 infrahilar region were noted(Fig. 1). Subsequent CT scan of the chest demonstrated multiple enlarged lymph nodes in the left hilar 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 hematoxylin-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 hereditary 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, Himmel-farb et al. (7) classified cardiopulmonary amyloidosis

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Received January 30, 1996; Accepted May 27, 1996

This article was supported by a research grant from the Catholic Medical Center

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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)

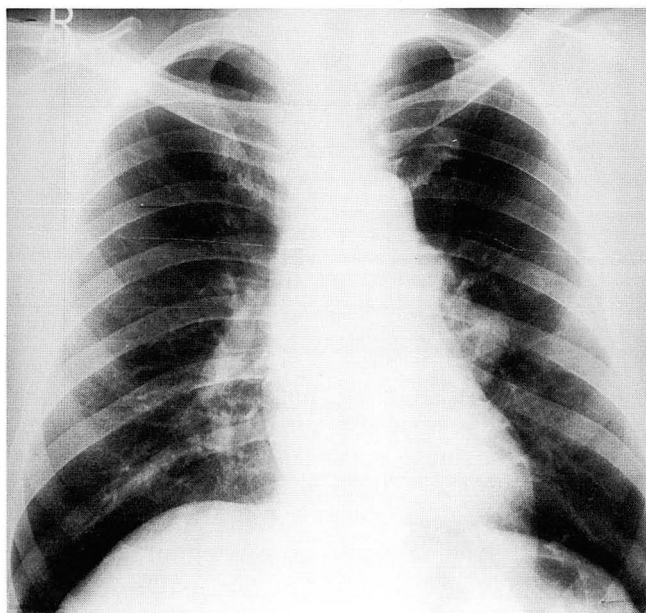


Fig. 1. A 55-year-old man with amyloidosis involving the mediastinal and hilar lymph nodes. Chest radiograph shows soft tissue masses in the left hilar and infrahilar regions, and the AP window. (AP : aortopulmonary)

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

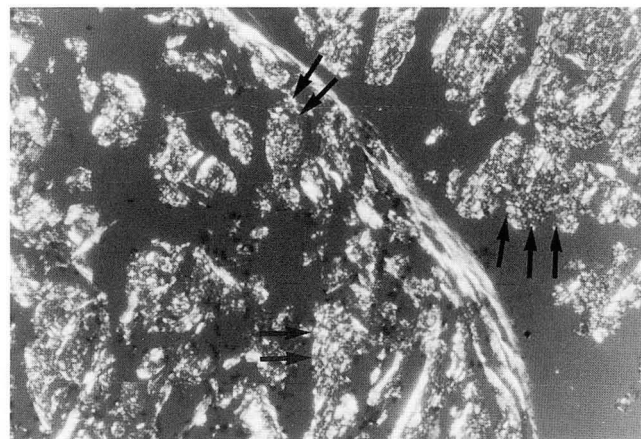


Fig. 3. Amyloid materials emitting birefringence. CT-guided biopsy specimen with Congo red staining shows virtual complete replacement of lymph node tissue by amyloid materials with typical apple-green birefringence(arrows) under the polarized light ($\times 100$).

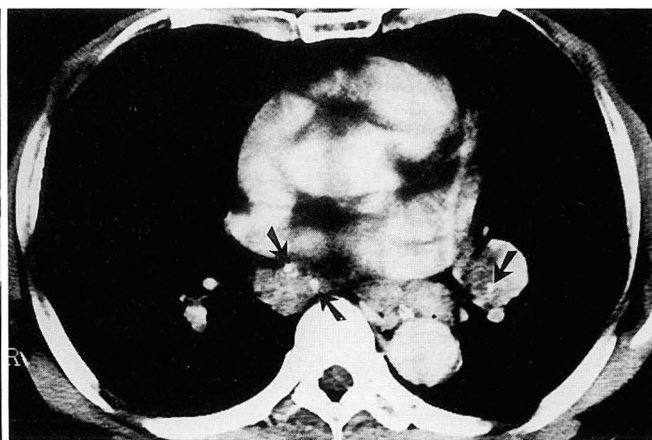


Fig. 2. Calcifications within the enlarged mediastinal and hilar lymph nodes.

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 azygosoesophageal recess, and the retrocardiac portion. Subcarinal lymph node is also enlarged with calcifications(not shown).

in cases with enlarged hilar or mediastinal lymph nodes, particularly in the presence of calcifications.

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대한방사선의학회지 1996; 34(5): 617~619

종격동과 폐문 임파절을 침범한 원발성 아밀로이드증: 1예 보고¹

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아밀로이드증은 인체내 여러 조직에 불용성 원섬유 단백질의 침착이 일어나는 드문 질환이다. 임파절을 침범한 아밀로이드증의 방사선학적 소견은 국내에서 보고된 바가 없다. 저자들은 종격동과 폐문 임파절을 침범한 아밀로이드증 1예의 단순 흉부 X선 사진과 전산화 단층촬영 소견을 보고하고자 한다.

제목 : Koala Sign

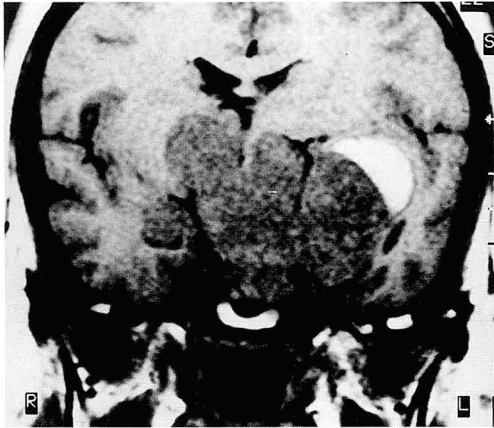


Fig. 1. Large pituitary adenoma on coronal T1WI

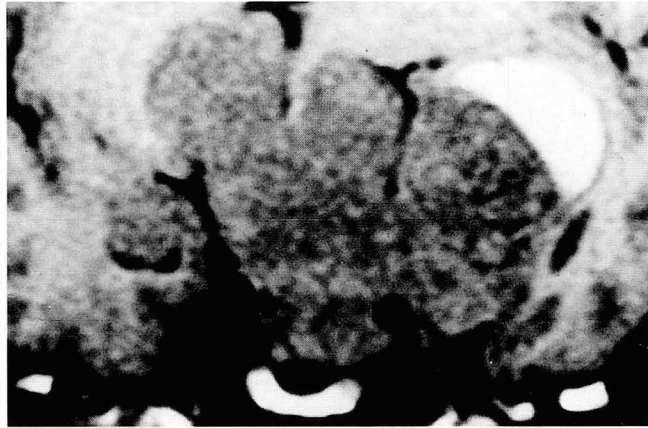


Fig. 2. Closed view of the mass

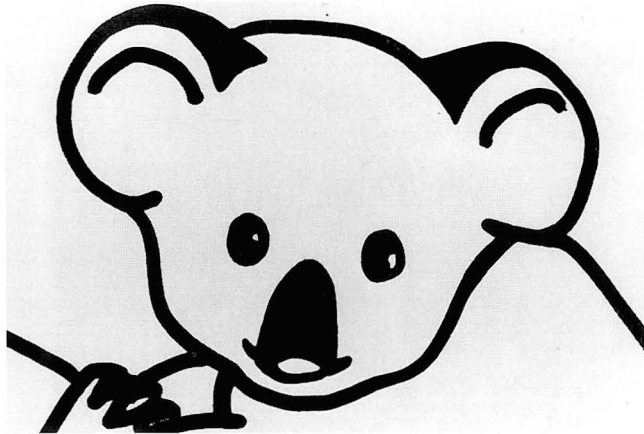


Fig. 3. Koala

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.

제공자 : 가톨릭대학교 의과대학 부천성가병원 진단방사선과
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