Demonstration of systemic lymphnodes by bone scintigraphy in amyloidosis

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We report a case of primary amyloidosis with calcification of systemic lymphnodes which were demonstrated as positives by bone scintigraphy. Positive sites delineated by bone scintigraphy would seem likely to reflect avid calcification of amyloid deposits. The discovery of positive systemic lymphnodes by bone scintigraphy is very rare in a routine study and may allow for amyloidosis in a differential diagnosis.

Key words: amyloidosis, calcification, lymphnode, bone scintigraphy

INTRODUCTION

BONE SCINTIGRAPHY is the commonest routine study of nuclear medicine. It can incidentally depict positive uptake in the soft tissue areas, most of which may be associated with calcification. Many disease states have been reported to show positive uptake of bone radioagents.1 However, demonstration of lymphnodes, particularly of systemic ones, by bone scintigraphy is very rare. In this paper, a case of primary amyloidosis in which systemic lymphnodes were delineated as positive will be presented.

A CASE REPORT

A 64-year-old man was admitted with complaints of persistent low-grade fever and coughs, with suspicion of pneumonia. He was already diagnosed as having primary amyloidosis 7 years ago by biopsies from inguinal lymphnodes, gastric and rectal mucosa, and by serological tests in another hospital. His complaints on the first admission were systemic lymphnode swelling and persistent fever. Since discharge, he has been followed at the Kitami Red Cross Hospital and has a clinical history of admission three times so far with gastric bleeding and pneumonia.

Received September 27, 1991, revision accepted December 6, 1991.

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On this admission, swollen lymphnodes were palpated bilaterally in symmetric sites of the posterior and anterior neck, axillae, and groin. In serological tests, serum M-protein (IgA\u00ed-type) was elevated and urine Bence-Jones protein negative, although urine protein was positive. The peripheral white blood cell count was 7,200/mm³, which is slightly elevated. No other abnormality was noted. A chest plain radiograph (Fig. 1) showed a radio-opaque area in the right lower lung and multiple nodular shadows in the mediastinal region bilaterally. The cardiac silhouette was also enlarged. A CT scan of the chest (Fig. 2A)

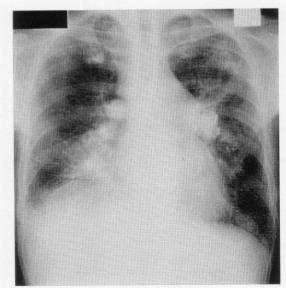


Fig. 1 A chest radiograph. Multiple calcified nodular shadows are observed in the mediastinal region.

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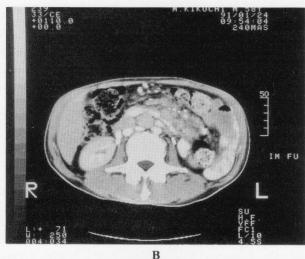


Fig. 2 Computed tomograms of the chest (A) and abdomen (B). Infiltrative calcified-density masses in the mediastinal region and pleural effusion are observed (A). In addition, an abdominal CT scan (B) shows multiple calcifications at sites of the para-aortic and mesenteric lymphnodes and massive deposits of calcified-density in the descending colon.

demonstrated infiltrative deposits of calcified density masses in the mediastinal region and pleural effusion bilaterally. In the abdomen (Fig. 2B), multiple calcifications were also revealed in regions of the paraaortic and mesenteric lymphnodes and massive calcified-density infiltration was observed in a part of the transverse and descending colon mucosa.

An electrocardiogram showed a low-voltage and complete right bundle branch block. Though the echocardiogram was normal, the patient was still suspected to have myocardial amyloidosis. Myocardial scintigraphy with ^{99 m}Tc-pyrophosphate performed showed multiple high uptakes of the radioagents in the hilar and mediastinal regions corresponding to calcified density masses on the CT scan (Fig. 3A). No

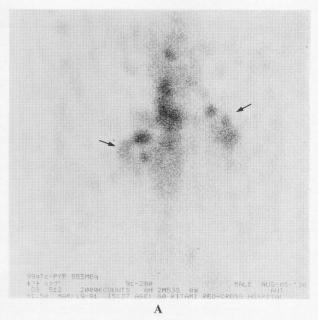




Fig. 3 Scintigrams with ^{99m}Tc-pyrophosphate (A) and ^{99m}Tc-HMDP (B). Both images demonstrate focal high uptakes in systemic lymphnodes (black arrows).

accumulation was revealed in the heart on either planar or SPECT images. In order to clarify general sites of calcification, whole body scintigraphy with ^{99 m}Tc-HMDP (Fig. 3B) was again carried out a few days later. An anterior whole body bone scintigram showed multiple uptakes in the systemic sites of

lymphnodes such as submandibular, right anterior neck, hilar, para-aortic, intra-pelvic and inguinal regions. No other uptake in organ-limited soft tissues was demonstrated. He died of pulmonary distress about 1.5 months after admission. An autopsy was not carried out.

DISCUSSION

Amyloidosis is a clinical manifestation of various disease states as well as a primary idiopathic condition, in which aggregates of proteincaneous fibrils are deposited in various body tissues.2 The main organ systems involved are the cardiovascular, gastrointestinal, genitourinary and nervous.3 Involvement of systemic lymphnodes is very rare. Clinical manifestations may be quite variable and the diagnoses obscure. The diagnosis was made with certainty by biopsy from various organ tissues. Non-invasive studies would be helpful not only for diagnosis but to evaluate the extent of the disease. Amyloid fibrils may be associated with osseous metaplasia.2 Therefore, bone scintigraphy has been reported to be positive in sites of amyloid deposits.4-8 However, positive delineation of sites of amyloid deposits by bone scintigraphy has not been high. It is reported to be 43% (3/7) with 99 mTc-Pyp and 18% (3/17) with 99 mTc-MDP.8 A mechanism for the high uptake of the bone radioagents is postulated to be related to the increased calcium content of the tissues involved rather than to the amyloid itself.8 In our case, scintigraphy demonstrated only systemic lymphnodes. although calcified area outside of the systemic lymphnodes were observed in colon mucosa by a CT scan. This phenomenon might be accounted for by the avidity of calcified masses to 99 mTc-Pyp or 99 mTc-HMDP. Positive delineation of sites of amyloid deposits by bone scintigraphy might be affected

by two factors: calcium contents and avidity of calcified masses to bone radioagents. In the latter contents, pyrophosphate is described to be more preferable for positive delineation of calcified amyloid than ^{99 m}Tc-MDP.⁸

In the view of differential diagnosis, demonstration of systemic lymphnodes by bone scintigraphy is very rare. Our case will allow us to evoke avid calcification of the lymphnodes as well as amyloidosis.

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