Detection of Bone Lesions by CT in POEMS Syndrome

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Abstract

Objective To study the utility of CT for detection of small bone lesions in POEMS (polyneuropathy, organomegaly, endocrinopathy, M-protein, and skin changes) syndrome. For patients with a solitary bone lesion, irradiation is a first-line treatment, whereas systemic chemotherapy is indicated for patients with multiple bone lesions. Therefore it is important to correctly identify the number of bone lesions.

Methods We studied the sensitivity of chest/abdomen/pelvic CT to detect bone lesions in 28 patients with POEMS syndrome. ^{99m}Tc-HMDP bone scintigraphy was performed in 14 patients, and the results were compared with CT.

Results CT showed multiple bone lesions in 68% of the 28 patients, and 71% of the lesions had a diameter <10 mm. In 14 patients who underwent both CT and scintigraphy, bone lesions were detected in 57% by CT and in 79% by scintigraphy, but the location and nature of the identified lesions were considerably different; CT frequently showed small lesions (diameter <10 mm) in the vertebrae and pelvis, which were not detected by scintigraphy, whereas scintigraphy could show lesions in the skull and long bones. Overall, by using both examinations, multiple bone lesions were found for 86% of patients.

Conclusion CT is particularly useful to detect small bone lesions. CT and bone scintigraphy are complementary, and therefore both should be performed for bone survey in POEMS syndrome.

Key words: POEMS syndrome, Crow-Fukase syndrome, sclerotic bone lesion, computerized tomography, CT, bone scintigraphy

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Introduction

POEMS syndrome (Crow-Fukase syndrome) is a rare systemic disorder associated with plasma cell dyscrasia and elevated serum vascular endothelial growth factor (VEGF). The acronym of this syndrome describes main clinical features: polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes (1-3). POEMS syndrome is a serious disease with neurological disability due to progressive polyneuropathy (4), and high mortality by multi-organ failure (5, 6). There are no randomized controlled trials in patients with POEMS syndrome (7), but several case reports and case series have shown benefits of treatments such as irradiation for

an osteosclerotic bone lesion (8), conventional chemotherapy (3, 6), high-dose chemotherapy with autologous peripheral blood stem cell transplantation (9-12), and thalidomide therapy (13).

Although the standard treatment for POEMS syndrome has not yet been established, it is generally recommended that for patients with a solitary bone lesion, focal irradiation is a first-line treatment, and patients who have multiple bone lesions or no detectable bone lesion should be treated with systemic therapy (7). Therefore, it is very important to correctly identify the number of bone lesions to determine the appropriate treatment. However, there is no systematic radiological study for bone survey in POEMS syndrome, presumably because of the rarity of the disorder. The aim of pre-

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Table 1. Bone Lesions Detected by CT in 28 Patients with POEMS Syndrome

Location*		
	Vertebrae	54%
	Pelvis	43%
	Sternum	11%
	Ribs/clavicles	21%
	Any of the above	68%
Nature**		
	Sclerotic	97%
	Lytic	1%
	Mixed	2%
Diameter**		
	< 10 mm	71%
	11 to 30 mm	23%
	30 mm <	6%

*percentage of 28 patients; **percentage of 146 lesions.

sent study was to investigate the utility of computerized tomography (CT) and scintigraphy for assessing bone lesions in patients with POEMS syndrome.

Subjects and Methods

Patients

This study included 28 patients with POEMS syndrome (20 men and 8 women). Their age ranged from 34 to 73 years (median, 48 years). All patients fulfilled diagnostic criteria of POEMS syndrome (7, 14). Twelve patients were examined before treatment, and the remaining 16 had received treatment with corticosteroids, melphalan, cyclophosphamide or thalidomide at the time of investigation.

Computed tomography and ^{99m}Tc-HMDP bone scintigraphy

Chest and abdominal CT (GE Medical System; Milwaukee, WI) were performed in all 28 patients. Scanning was made from the upper margin of the sternum to the ischium. The slice thickness was 10 mm. Benign bone lesions (bone island, hemangioma and Schmorl's node) were excluded, and bone lesions were classified into three groups; osteosclerotic, osteolytic and mixed lesions. Osteosclerotic lesion was defined as a lesion with high density surrounded by normal bone marrow or fused with bone cortex, whereas osteolytic lesion was defined as a lesion with low density in the bone cortex.

Bone scintigraphy was performed in 14 patients using an intravenous injection of 555 MBq of ^{99m}Tc-HMDP. Whole-body planar images were taken 4 h after injection using dual-headed gamma camera. The images were displayed 256×1024 matrix and reported by radiologist. Lesions were identified as focal tracer uptake. For both CT and scintigraphy, the abnormalities were judged by a radiologist (T.H.) who was blinded to clinical information.

Results

Computed tomography

Table 1 shows location, nature, and size of bone lesions on chest and abdominal CT. Of the 28 patients with POEMS syndrome, 19 (68%) had multiple bone lesions. The lesions were frequently found in the vertebrae and pelvis. The total number of abnormal bone lesions in patients with POEMS syndrome was 146, and the large majority of lesions was sclerotic (97%) and the diameter was 10 mm or smaller (71%). Representative multiple osteosclerotic lesions in the vertebra and pelvis are shown in Fig. 1.

Comparison of CT and ^{99m}Tc-HMDP bone scintigraphy

Table 2 shows comparison of findings on CT and scintigram in 14 patients with POEMS syndrome. For lesions located in the vertebrae and pelvis, the sensitivity is higher on CT than scintigraphy. Small bone lesions found on CT could not be detected by the scintigram frequently (Fig. 1). By contrast, the sensitivity was higher with scintigraphy for the rib/clavicle lesions, and scintigraphy detected lesions in the skull in 21% of the patients, and in the long bones in 7%. Using both CT and scintigraphy, 86% of the patients had bone lesions, and 50% had bone lesion in the vertebrae. Of the 14 patients, 12 (86%) had multiple lesions, whereas the remaining two (14%) had a single lesion.

Discussion

Our results show that CT and bone scintigraphy are useful tools to detect bone lesions in patients with POEMS syndrome. CT is particularly sensitive to small lesions with a diameter <10 mm in the vertebrae and pelvis, whereas scintigraphy can detect lesions in the skull and long bone. In addition, our CT findings confirmed that almost all of the bone lesions are osteosclerotic and multiple in POEMS syndrome. Using both CT and scintigraphy, 86% of the patients examined had multiple bone lesions.

Using plain X-ray of skull, vertebrae, pelvis, femur, and humerus, previous studies have shown bone lesions in 55% of 104 patients (5), 50% of eight patients (15), and 27% of 99 patients (16). The sensitivities are significantly lower than that of our study (86%), and it is not practical to examine each part of the whole body by plain X-ray. These studies showed that bone lesions were most frequently in the vertebrae and pelvis, and our study confirmed the findings. A recent study showed that in one of four patients with POEMS syndrome, FDG-PET can detect a pelvic lesion that is not detected by other imaging studies (17). The utility of FDG-PET should be studied in future studies.

Nakanishi et al (5) revealed that histological examination showed plasmacytoma in most of the osteosclerotic lesions in patients with POEMS syndrome. Our results confirmed

Figure 1. Findings of vertebral and pelvic CT and ^{99m}Tc -HMDP skeletal scintigraphy in a 64-year-old woman with POEMS syndrome. Multiple small osteosclerotic lesions in the lumbar vertebra (A) and pelvis (B) are detected on CT. Scintigraphy shows focal tracer uptake in the ribs (arrows), but fails to show lesions in the vertebrae and pelvis (C).

Table 2. CT and Bone Scintigraphy Findings in 14 Patients with POEMS Syndrome

	CT	Bone scintigraphy	Any of the two
Vertebrae	50%	36%	50%
Pelvis	36%	14%	43%
Sternum	7%	7%	14%
Ribs/clavicles	14%	36%	36%
Skull	Out of the field	21%	21%
Long bones	Out of the field	7%	7%
Any of the above	57%	79%	86%

that 97% of bone lesions detected by CT were osteosclerotic. Therefore we speculate that almost all of the bone lesions are plasmacytomas in this study. Current diagnostic criteria for POEMS syndrome includes sclerotic bone lesion as a major criterion (7, 14). Previous studies and this study showed that most of bone lesions are osteosclerotic (5, 15, 16). The present study showed that by using both CT and scintigraphy the sensitivity to detect bone lesions could be increased, and therefore the use of both techniques would contribute to the diagnosis.

In this study, the sensitivity was 57% by CT and in 79% by scintigraphy, but location and nature of the identified lesions were considerably different in the two imaging tools; CT frequently showed small lesions (diameter <10 mm) in the vertebrae and pelvis, which were not detected by scintigraphy as shown in Fig. 1, whereas scintigraphy has advan-

tage to evaluate the whole body, and could show lesions in the skull and long bones. We suggest that both CT and scintigraphy should be performed for bone survey in POEMS syndrome, and this would contribute to the diagnosis and appropriate treatment in patients with POEMS syndrome.

The authors state that they have no Conflict of Interest (COI).

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