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EVALUATION OF SKELETAL MUSCLE WITH THALLIUM-201 SCINTIGRAPHY IN MYOTONIC MUSCULAR DYSTROPHY: A CASE REPORT

SHUHEI YAMAMOTO, M.D.

First Department of Internal Medicine, Nagoya University School of Medicine, Nagoya 466, Japan

ABSTRACT

A case of myotonic muscular dystrophy in which thallium-201 showed dystrophic change of skeletal muscle is described. After i.v. injection of 2 mCi of thallium-201, the patient's whole body scintigram was reconstructed by taking the geometric mean of the corresponding anterior and posterior pixel counts over the 512 × 512 matrix format. The scintigram showed manifest hypoperfusion at the site of muscle weakness. Thallium-201 whole body scintigraphy was thought to be useful for noninvasive evaluation of skeletal muscular involvement in muscle disease.

Key Words: Thallium-201, Whole body scintigraphy, Myotonic dystrophy, Skeletal muscular involvement

A case of myotonic muscular dystrophy in which thallium-201 leg scan was useful for evaluation of dystrophic change of skeletal muscle is described. This case is, to the author's knowledge, the first reported case of skeletal muscular dystrophy in which the disease was also demonstrated with ^{201}Tl.

CASE REPORT

A 36-year-old male had slight muscle weakness of left leg. The extremities were symmetrical with no noticeable muscle atrophy, fasciculation, edema, or deformity of the joints (Fig. 1). Both legs were warm, showing good pulsations of the dorsalis pedis arteries. There was no limitation of movement in any joints. Slight left calf muscle weakness was found, and left Achilles tendon reflex was absent. The patient could walk freely and clinical stage according to Swinyard & Deaver was 1, i.e., the mildest one.

Myotonia was elicited by percussing the thenar eminence. No signs of cranial nerves, alopecia, cataract, sensory disturbance, testicular atrophy, or mental impairment were found. There was no history or symptoms of peripheral vascular diseases. No specific therapy, such as drugs, exercises, splints, braces, or corrective surgery, has been done as yet.

Electromyograms of both legs showed the characteristic afterpotentials of myotonia together with the usual myopathic pattern of many low-voltage, brief action potentials, confirming the diagnosis.

Laboratory findings showed high serum CK level (100–400 i.u./ml) and creatinuria. Complete blood counts, serum electrolytes, liver function test, levels of IgG, parathyroid and thyroid hormones, glycosuria, proteinuria, and serologic test for syphilis were normal or negative.

山本秀平
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This patient was referred to the author for evaluation of left ventricular fibrosis suspected from the abnormal standard 12-lead ECG. Scanning of the whole body was performed immediately after myocardial imaging.

METHOD

Whole body scintigraphy was performed with a rotating digital gamma camera with twin opposing, large rectangular detectors (Toshiba GCA 70A TOKU) and on-line computer (Toshiba GMS 55A) after intravenous injection of 2 mCi of thallium-201. The effective field of view of the two detectors was 35 cm by 50 cm. Anterior and posterior data of the whole body were acquired simultaneously with a 512 by 512 matrix by the twin opposing detectors.

The two cameras scanned the whole body from head to feet in 15 min. Photon attenuation of posterior data through the scanning table was corrected using the theoretical value gained by phantom experiment. Whole body scintigram of thallium-201 was reconstructed by taking the geometric mean of the corresponding pixel values of the two data. Region of interest (ROI) was

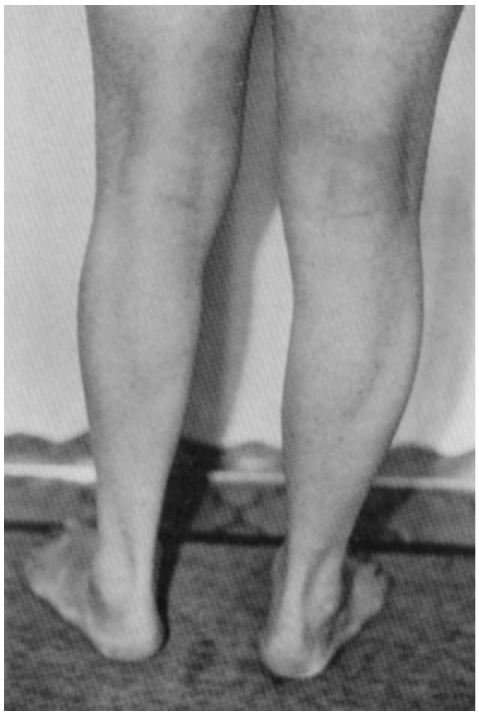

Fig. 1

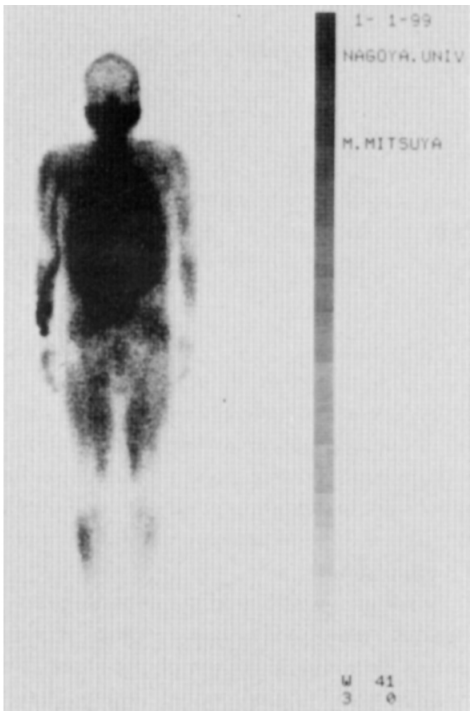

Fig. 2

Fig. 1 Photograph of the patient's legs.
Note that there is no muscle atrophy in either of the calf muscles.

Fig. 2 Whole body scintigram of the patient.
Twenty minutes after i.v. injection of thallium-201, whole body scintigraphy was performed using twin opposing, large rectangular detectors. Thallium-201 is distributed mainly to the parotid glands, thyroid glands, heart, liver, and kidneys. It is distributed mildly to skeletal muscles, lungs and visceral organs. Note that distinct thallous hypoperfusion is demonstrated at the left calf muscles.

defined manually over each organ. In the legs, ROIs were located over the thighs and calves symmetrically, and the mean pixel count of each ROI was calculated.

RESULTS

The reconstructed image of whole body distribution of thallium-201 showed selective loss of uptake in the left calf muscles and residual venous radiotracer activity at the injection site of the right arm (Fig. 2). Thallium-201 also accumulated symmetrically in the bilateral scapular, deltoid, and gluteal muscles.

Mean pixel counts of ROI placed on each part of limbs in the whole body scintigram were as follows: right thigh, 20.3 ± 8.3; left thigh, 19.6 ± 7.9; right calf, 11.9 ± 6.2; left calf, 7.3 ± 3.9, as measured 20 min after i.v. injection of ^{201}Tl (Table 1).

Based on the fact that both legs were warm and the bilateral dorsalis pedis arteries showed good pulsations, laterality in leg blood flow was not thought to be present.

DISCUSSION

Of the available radioactive tracers today, thallium appears to be one of the most suitable tracers for evaluation of organ perfusion. It disappears extremely rapidly from circulation following intravenous administration, and is immediately deposited intracellularly with a distribution pattern depending mainly on the nutrient blood flow to the organ, the tissue permeability to the thallous ion, and the activity of the sodium-potassium adenosine triphosphatase transport system.

Thallium-201 myocardial imaging is used in evaluating infarction at rest or ischemia at peak exercise in coronary artery disease. It is also used in assessing fibrosis or degeneration in various types of myocardial diseases, including muscular dystrophy.

Thallium-201 leg scan has been used clinically in detecting and localizing peripheral arterial

Table 1. Thallium counts in various parts of the body.

Organ		No. of Pixels	Maximum	Average	S.D.
Thyroid gland		248	81	56.7	9.99
Shoulder	(right)	364	49	29.8	7.78
	(left)	360	49	28.4	6.88
Deltoid muscle	(right)	364	49	29.4	8.49
	(left)	452	30	19.1	6.10
Heart		312	156	98.7	19.18
Lung	(right)	460	90	55.3	12.53
	(left)	420	72	43.6	9.08
Liver		796	132	85.1	21.98
Abdomen		4312	104	34.9	14.18
Kidney	(right)	380	225	134.9	31.74
	(left)	404	169	100.5	31.01
Thigh	(right)	2108	42	20.3	8.34
	(left)	1980	42	19.6	7.89
Calf	(right)	1104	30	11.9	6.23
	(left)	1216	20	7.3	3.94

diseases since the 1970s, as perfusion defect is known to occur in diseases causing leg ischemia at its peak exercise.[1,2]

However, there have been as yet few reports on skeletal muscle diseases diagnosed with radioactive tracers, e.g., a case of dermatomyositis with thallium perfusion defect induced by bicycle ergometer,[3] a case of unilateral tremor with abnormal accumulation of thallium at the diseased extremities,[4] and a case of a marathon runner with accumulation of technetium-99m at site of rhabdomyolysis.[5]

Nutrient blood flow of skeletal muscles averages 3 to 4 ml per min per 100 g of muscle. Uniform thallous radiotracer-activity expressing muscle contour is observed in a normal case. Both being striated muscles, skeletal and cardiac muscles have similar physiologic properties, and in muscular dystrophy, similar pathologic change as well.

It is, therefore, reasonable to assume that thallium uptake would decrease if skeletal muscle is dystrophic, and would increase if hypertrophic, as it does in myocardium. A result which supports this assumption was obtained in the case presented in this paper. Site of skeletal muscular dystrophy coincided with the results of thallium-201 whole body scintigraphy. Although imaging of the patient was performed 20 min after injection of thallium-201 according to the methods prescribed before,[3,4] little is known about the biological half-life of thallium in various muscle diseases. Therefore, the most appropriate time for muscle imaging remains to be solved by future investigation.

Besides the method presented in this paper being useful for early detection of muscular dystrophy, it enables evaluation of skeletal involvement without exercise stress test, which cannot be easily performed in an advanced condition. Indeed, electromyography, muscle biopsy, and X-ray CT are useful for evaluating fibrosis and fatty infiltration of skeletal muscles in muscular dystrophy, but these methods have some shortcomings in that whole muscles of the four extremities cannot be assessed at one time. A drawback of thallium-201 scintigraphy, however, is that evaluation of arm used for ^{201}Tl injection may be inevitably difficult when strong radiotracer activity remains at that site.

Not only myotonic muscular dystrophy, a rare disorder occurring in about one in 100,000 people in Japan, but also myopathies due to other systemic diseases may be evaluated with thallium-201. Thallium limb scan is noninvasive, simple to perform, and not time-consuming. Thus, the method may be useful for screening the extent of skeletal muscular disorder. In addition, the equipment used has two unique aspects: first, data can be acquired from anterior and posterior aspects simultaneously with the two opposing detectors; and second, whole body scintigraphy can be easily performed at one time using the rectangular detectors with a large field of view. This attractive imaging procedure seems to open up a new field in the area of diagnostic myology.

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