COMPUTED TOMOGRAPHY OF SKELETAL MUSCLES IN
CHILDHOOD SPINAL PROGRESSIVE
MUSCULAR ATROPHIES

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Computed tomographic (CT) scanning of skeletal muscles was performed in patients
with type 1 and type 2 spinal progressive muscular atrophy (SPMA) and Kugelberg-
Welander disease (K-W) to delineate the characteristic CT features of each category.
Marked muscular atrophy was observed in type 1 SPMA, and both muscular atrophy and
intramuscular low density areas in type 2 SPMA, changes being more pronounced in
older patients. In contrast, in K-W, muscular atrophy was slight, and intramuscular low
density areas constituted the most prominent findings. These observations indicate that
SPMA and K-M are each characterized by distinct CT findings.

Introduction

Computed tomography (CT) of skeletal muscles has been widely applied to various neuromuscular
disorders since 1977, when O'Doherty and his colleagues reported CT changes in cases of
pseudohypertrophic muscular dystrophy11, and the utility of the technique is presently being
established as a non-invasive means of in situ evaluation of muscle involvement. In muscular
dystrophy, a characteristic CT pattern of muscle involvement has been confirmed. In addition to
allowing comparison of severity of involvement of different muscles within a single individual, CT
diagnosis also serves to distinguish primary from neurogenic. However, recent reports of muscle CT
studies are not sufficient to determined individual CT patterns in various forms of neurogenic. In the
present study, we investigated skeletal muscle CT scans in cases of spinal progressive muscular
atrophy (SPMA) in order to identify characteristic CT features of SPMA.

Patients and Methods

The patients included one case of type 1 SPMA (age 11 months), four cases of type 2 SPMA (ages
1–6 years), and one case of Kugelberg-Welander (K-W) disease (age 13 years). The diagnosis had
been established in each case by means of a comprehensive work-up including neurological,
electrophysiological and biochemical examinations as well as muscle biopsy. Clinical profiles of
the patients, including motor ability at the time of the CT study, are shown in Table. CT examina-

<table>
<thead>
<tr>
<th>Type of disease</th>
<th>Age (y. m)</th>
<th>Sex</th>
<th>Motor ability</th>
</tr>
</thead>
<tbody>
<tr>
<td>SPMA type 1</td>
<td></td>
<td></td>
<td>Bedridden (under respiratory assistance)</td>
</tr>
<tr>
<td>type 2</td>
<td>1 : 4</td>
<td>M</td>
<td>Turning around while sitting</td>
</tr>
<tr>
<td></td>
<td>4 : 2</td>
<td>F</td>
<td>Turning around while sitting</td>
</tr>
<tr>
<td></td>
<td>6 : 2</td>
<td>M</td>
<td>To keep sitting with support</td>
</tr>
<tr>
<td></td>
<td>6 : 9</td>
<td>M</td>
<td>To keep sitting with support</td>
</tr>
<tr>
<td>K-W</td>
<td>13 : 0</td>
<td>F</td>
<td>Ambulatory with assistance</td>
</tr>
</tbody>
</table>

SPMA : Spinal progressive muscular atrophy
K-W : Kugelberg-Welander disease
Fig. 1 Normal muscle CT scans and innervated spinal level of each muscle
From the top, horizontal slices at the level of the 3rd lumbar vertebra, mid-thigh and the portion of the calf having the largest diameter.

Results

The images in the control subject were confluent, not sharply demarcated, and homogeneous in density. In the patient with type 1 SPMA (Fig. 2), all muscles at each slice level displayed overall reduction in the size of the muscle itself surrounded by large areas of low density, and an increased separation between the individual muscles. These changes represent marked muscle atrophy on CT. Furthermore, the paravertebral muscles, in particular, exhibited almost entirely low density (LD) lesions, which are considered to be a result of pronounced muscle atrophy with replacement of muscles by fatty tissue. However,
distinct LD areas were not found within other muscles. In the patients with type 2 SPMA (Fig. 3), atrophic changes were noted in all muscles, although to a lesser degree in comparison to those in the type 1 SPMA case (Fig. 3A, B). Atrophic changes became more pronounced with increasing age, and moth-eaten LD areas were noted within

![Muscle CT scans of type 2 spinal progressive muscular atrophy (SPMA)](image)

**Fig. 3** Muscle CT scans of type 2 spinal progressive muscular atrophy (SPMA)
Atrophy of all muscles was recognized, although not to the same extent as in the type 1 SPMA case. Atrophic change progressed with increasing age. Furthermore, low density areas were noted within muscles, particularly pronounced in the triceps muscle of the calf. (A; 1 year of age, B; 4 years of age, C, D; 6 years of age)
Fig. 4 Muscle CT scans of Kugelberg-Welander disease
Muscular atrophy was slight, but intramuscular low density changes were conspicuous. The degree of involvement was variable even in contiguous muscles, thus the findings were similar to those ordinarily seen in muscular dystrophy.

In the patient with K-W disease (Fig. 4), only slight muscular atrophy was detected, but moth-eaten or striate LD changes were conspicuous at each level. In particular, the paravertebral muscles and the quadratus lumborum displayed almost complete LD images. In the thighs, the rectus femoris muscle was well preserved, but a marked LD area extended over a wide range in the vastus; in the gracilis and sartorius, changes apparently corresponding to the compensatory hypertrophy seen in muscular dystrophy were observed. In the calves, the soleus showed a distinctive LD image within the muscle whereas the other muscles were relatively well preserved.

The skeletal muscle CT patterns of SPMA

Figure 5 shows a schematic diagram of the observed CT changes in SPMA and in K-W. Severe muscular atrophy was observed in type 1 SPMA, and muscular atrophy as well as LD regions in type 2 SPMA. These changes were especially pronounced in cases of long duration in this type of SPMA. On the other hand, in K-W, muscular atrophy was slight, and intramuscular LD areas constituted the most prominent finding. Moreover, the degree of involvement varied even within the same effector muscle group which is in agreement with characteristic findings in muscular dystrophy.

Discussion

According to the general view, the principal muscular CT changes in neurogenic disorders are atrophy, while LD regions emerge concomitantly with progression of the disorder. As regards muscle CT images in type 1 SPMA and K-W, Hawley et al. reported that type 1 SPMA displays muscular atrophy typical of neurogenic disorders, while K-W showed CT changes extremely similar to those of muscular dystrophy. In particular, in a case of K-W, Hawley et al. noted different changes even in contiguous muscles, such as those observed in Duchenne type muscular dystrophy, and for the first time proposed that CT findings in K-W differ from those of disorders originating in other anterior horn cell diseases. Neuropathologically,
type 1 SPMA is known to be characterized by uniform loss and degeneration of the neurons of the anterior horns, except for those of cervical nerves C3-C5, which innervate the diaphragm\(^3\). In the present study, also, the muscles innervated by motor neurons below the L1 level showed uniform atrophic changes. In K-W, however, changes are known to extend over all levels of the spinal cord\(^4\text{-}^6\), and discrepancies with the characteristics of CT images have been noted. For example, even if the paravertebral muscles, also innervated by L3 neurons, are completely replaced by fat, the rectus femoris, gracilis, and sartorius are well preserved (Fig. 1). However, these discrepant findings may be explained by taking muscle pathology into consideration. That is, in type 1 SPMA, the histopathological picture is characterized by the presence of hypertrophic fibers which are sporadically distributed within large groups of atrophic fibers, whereas in K-W, atrophic fibers tend rather to form small aggregates, or small angulated fibers are sporadically present, and myogenic changes such as degeneration or necrosis of muscle fibers and proliferation of connective tissue, are observed at these sites. The pathogenesis of these additional myogenic changes in K-W remains obscure. In any case, the CT scans in the present study reconfirmed that SPMA and K-W show different manifestations according to histological background.

As regards type 2 SPMA, in 1986, Nagao et al\(^7\) reported on CT findings of young children whose ages ranged from one to three years, while Hori-kawa et al\(^8\) reported on CT findings in older cases ranging in age from six to 22 years; according to both reports, the CT changes consisted of muscular atrophy and intramuscular LD areas. The muscular pathology observed in type 2 SPMA is regarded as essentially identical with that of type 1 SPMA, but since the clinical course is more gradual, reinnervation of denervated muscles can be assumed to occur. Consequently, muscular atrophy in type 2 SPMA does not progress as abruptly as that in type 1 SPMA. Moreover, in muscles with chronically progressive atrophy, the atrophied fascicles are replaced by connective or adipose tissue, and this presumably is responsible for the appearance of LD areas on CT scans. Furthermore, the results obtained in the present study also agree with those obtained by Nagao et al\(^7\), that is the triceps muscle of the calf particularly tended to show more severe atrophy than the thigh muscles; this is an extremely interesting finding in SPMA, which is generally characterized by predominantly proximal muscle weakness.

The results of the present study suggest that skeletal CT scans provide images which faithfully reflect the myopathic picture in SPMA as well as other neurogenic disorders, and constitute a highly effective means of examination which permits accurate assessment of the degree of muscular lesion severity.

References

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小児脊髄性進行性筋萎縮症の骨格筋 CT スキャン

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小児脊髄性進行性筋萎縮症（SPMA）I型、II型および、Kugelberg-Welander 病（K-W）に骨格筋 CT スキャンを施行し、各臨床型における筋 CT 上変化の特徴を評価検討した。SPMA I型 1例（11ヶ月）、II型 4例（1 〜 6歳）および K-W 1例（13歳）を対象にし、第2腰椎（L3）、大腿部および下腿最大周径レベルをスライスし、各筋の CT 画像を肉眼的に評価した。SPMA I型では各レベルの全ての筋で著しい萎縮を認めた。他の筋の筋内 LD は目立たなかった。SPMA II型では、I型ほどではないがすべての筋に萎縮を認め、特に各筋内に虫喰い状低吸収域（LD）が認められた。筋萎縮は年長例ほど強く、また LD 出現も著明であった。一方、K-W においては、各レベルとも筋萎縮はわずかに認められるのみで筋内の虫喰い状または縦状 LD の変化が著明であった。また、大腿において筋ジストロフィー症に認められるような筋の選択的罹患性や、一部の筋に代償性肥大と思われ変化を認めた。即ち、SPMA I型では筋の萎縮、II型では筋萎縮と筋内 LD を、また K-W では筋萎縮よりも筋内 LD が変化の主体であり筋ジストロフィー症の特徴的所見と近似していた。これら結果から、SPMA I型、II型と K-W の筋 CT 上異なる特徴を示唆された。

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