ULTRASTRUCTURAL, HISTOCHEMICAL AND IMMUNOHISTOCHEMICAL STUDIES ON MUSCLE LESIONS OF MYOTONIC DYSTROPHY-LIKE MYOPATHY IN A MUTANT LWC STRAIN OF JAPANESE QUAILS

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INFORMATION

Hokkaido University granted the degree of Doctor of Veterinary Medical Science (equivalent to Ph. D.) to the following 1 graduate of the Graduate School of Veterinary Medicine on September 30, 1996.

The title of the thesis and other information are as follows:

ULTRASTRUCTURAL, HISTOCHEMICAL AND IMMUNOHISTOCHEMICAL STUDIES ON MUSCLE LESIONS OF MYOTONIC DYSTROPHY-LIKE MYOPATHY IN A MUTANT LWC STRAIN OF JAPANESE QUAILS

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A newly established strain of Japanese quails (LWC strain) has histologically muscle lesions similar to those in the myotonic dystrophy of man: ring fibers, sarcoplasmic masses and atrophy in types 2A and 2B fibers. This strain is inherited as an autosomal dominant trait and is homozygous lethal. The affected birds were incapable of lifting their wings vertically upward (wing lift test) and were unable to right themselves when placed on their dorsum (flip test). Electromyographic examination of their muscles revealed a myotonic discharge. In the present study, muscles in the LWC quails were investigated ultrastructurally, histochemically and immunohistochemically.

Ultrastructurally, the ring fibers were composed of myofibrils with distinct A and I bands and M and Z lines, although these bands and Z-lines were sometimes indistinct. The myofibrils run perpendicularly between the normal myofibrils and plasma membrane. The sarcoplasmic masses were also found between the normal myofibrils and plasma membrane, which consisted of fragmented myofibrils, triads, mitochondria, glycogen granules and ribosomes, among which there was no systemic interconnection. In the atrophic fibers, myofibrils were partially destructed and replaced by autophagic vacuoles, lysosomes, glycogen granules, mitochondria with dense matrix and cristae, and honeycomb structures. The plasma membrane of the atrophic fibers had multiple projections and folds. Occasionally the atrophic fibers were surrounded by a redundant basement membrane. The honeycomb structures might be derived from a proliferation of transverse tubules and were observed frequently in the atrophic fibers.
and occasionally in the apparently normal fibers in LWC quails.

From the findings obtained here, it was suggested that one of the possible causes of the ultrastructural changes in the muscles of LWC quails is a defect in the cytoskeleton controlling the cellular organelles.

In the normal quails, muscle fibers were of three major types, types 1, 2A and 2B histochemically. In the superficial part of the muscles examined, type 2A fibers occupied the inner area of the fascicles and type 2B fibers were found at the rim of the fascicles. Immunohistochemically, type 2A fibers reacted positively with the anti-adult fast MHC antibody, while type 2B fibers were only weakly labeled with it. Type 1 fibers showed a strong positive reaction with the anti-slow MHC antibody but did not react with the anti-adult fast MHC antibody. With the anti-N-CAM antibody, the muscle fibers were labeled consistently negatively, except for a few cells including some satellite cells. In the superficial fascicle of the muscle examined in LWC quails, type 2A fibers showed reduced stainability with the myofibrillar ATPase and anti-adult fast MHC antibody. On the contrary, type 2B fibers revealed increased immunoreactivity for both ATPase and anti-adult fast MHC antibody and, were also reduced in diameter. No changes were detected in the deep fascicle of the muscle. The sarcoplasmic masses were labeled with both anti-adult fast and slow MHC antibodies. A number of muscle fibers were positive with the anti-N-CAM antibody, which were distributed in a random fashion throughout the muscle except the deep fascicle. The intensity of the labeling varied markedly from fiber to fiber. The ring fibers and sarcoplasmic masses were also reacted positively with the anti-N-CAM antibody. However, there was no particular association between the N-CAM positive fibers and ring fibers or sarcoplasmic masses.

From these findings, it was suggested that the sarcoplasmic mass was formed as a sequel of muscle fiber hypertrophy occurring with defects in the cytoskeleton. Moreover, morphological, histochemical and immunohistochemical changes in both types, 2A and 2B fibers might have been induced by long-term continuous contraction of muscle fibers resulting from myotonia.
