[PICTURES IN CLINICAL MEDICINE]

Dropped Head Syndrome Caused by Immune-mediated Necrotizing Myopathy

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Key words: dropped head syndrome, immune-mediated necrotizing myopathy, anti-SRP antibodies, HyperCKemia, idiopathic inflammatory myopathies

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Picture 1.

Picture 2.

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Picture 3.

The patient was a 60-year-old Japanese woman who presented with dropped head syndrome (DHS) and limb weakness (Picture 1). A neurological examination showed proximal dominant limb and truncal weakness. The neck extensor strength was weaker than the neck flexor strength. She had increased creatine kinase levels (8,430 U/L) and anti-signal recognition particle antibodies. Short-tau inversion recovery images showed paraspinal muscle hyperintensity (Picture 1). Hematoxylin-eosin staining of a left biceps muscle specimen showed mild-to-moderate fiber size variation, and necrotic and degenerating processes without mononuclear cell infiltration (Picture 2). Immunostaining for membrane attack complex was positive in the sarcolemma of necrotic fibers (Picture 2). The results of the histopathological evaluation were compatible with immune-mediated necrotizing myopathy (IMNM). DHS and the short-tau inversion recovery abnormality improved after intravenous methylprednisolone pulse therapy followed by treatment with oral prednisolone and tacrolimus (Picture 3).

DHS is characterized by severe neck extensor weakness and occurs in various neuromuscular disorders (1). Although

neck weakness is found in IMNM, the association of DHS is unknown (2). Clinicians should consider IMNM if patients with DHS have hyper-creatine kinase-emia.

The patient provided her written informed consent for the publication of this report.

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

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