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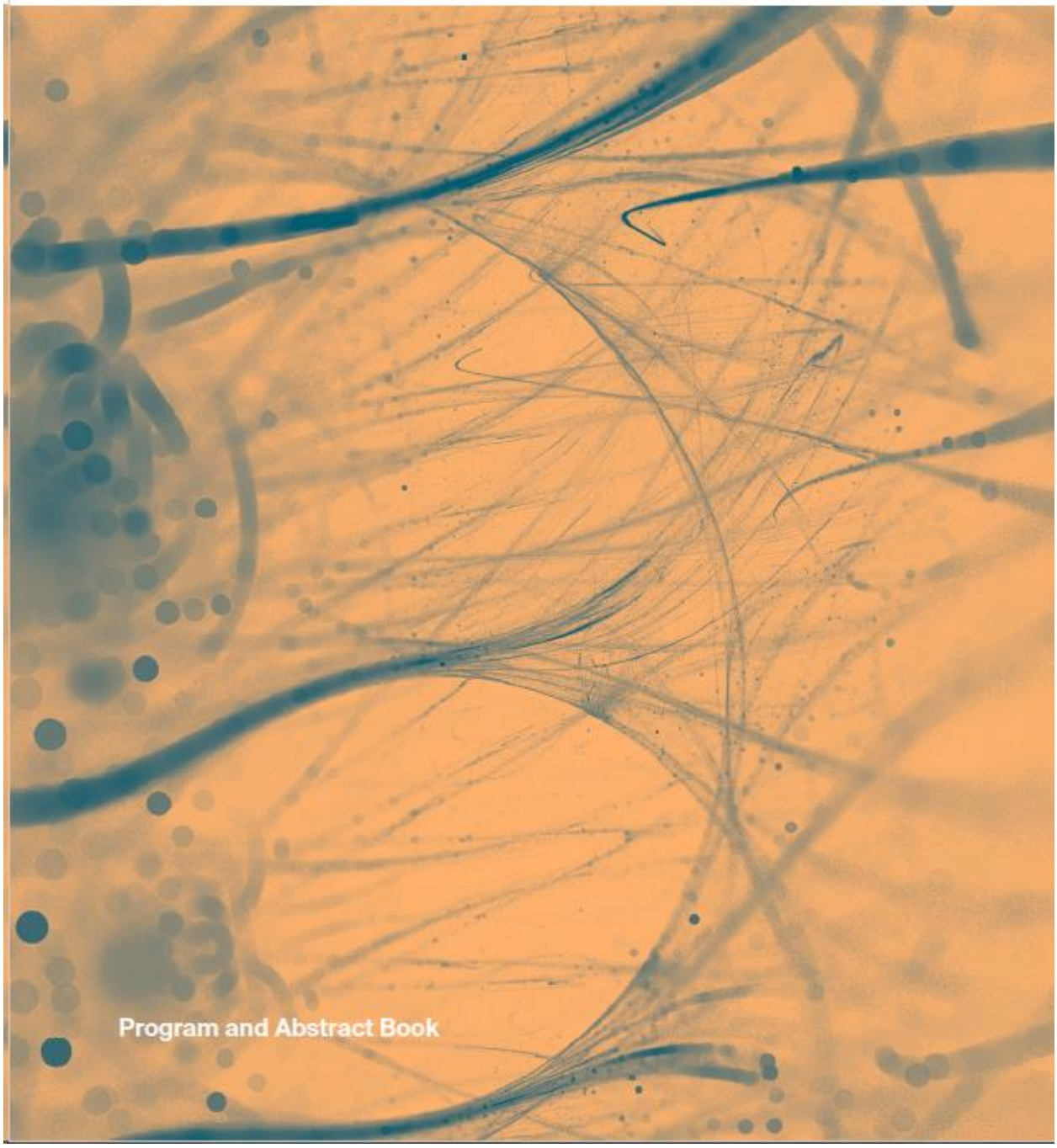

sciforum-098838: Monomyelic amyotrophy. A clinical case

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Introduction

Monomelic amyotrophy (Hirayama disease) is a rare form of non-progressive motoneuron disease caused by necrotic lesions of the anterior horns of the spinal cord. Purpose of the study. To evaluate the condition of the patient's leg muscles by electromyography.

Methods

A 17-year-old young man with complaints of weakness and fatigue in both hands (more pronounced on the right side) and weakness in legs when bending the head was examined. Clinical and neurological, electrophysiologic analysis, MRI of the head and neck while performing functional tests were performed. To assess muscle strength, the patient performed maximal dorsal flexion of the foot on himself, head straight and head down, while EMG was recorded from the anterior tibial muscles of both legs.

Results

MRI data revealed characteristic signs of monomyelic atrophy. Atrophy of the hand muscles on the right side, fascializations were clinically revealed. During neck flexion, there was weakness 4b in the extensors of the feet after 1 minute. The amplitude of the motor response of the tibialis anterior muscle during head tilt on the right decreased by 4 times, and on the left by 6 times, with a more pronounced decrease in strength in the upper extremities in the right arm than in the left. According to the literature, the "classical" variant of monomyelic amyotrophy is not characterized by sensory disorders, pyramidal symptoms, and involvement of leg muscles.

Conclusions

The use of electrophysiological methods of investigation along with neuroimaging methods allowed to establish a more accurate picture of the course of the disease.