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Distribution of Some Extracellular Matrix Proteins and Ultrastructural Findings in Sural Nerve Biopsy in Demyelinating Disease

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Abstract: The involvement of both the peripheral nervous system (PNS) and central nervous system (CNS) in multiple sclerosis is a seldom encountered combination in neurology clinics. In this report, immunohistochemical and ultrastructural techniques were used to analyse the disease course of a patient exhibiting involvement of both systems. In 1980, a 44-year-old man was admitted to our clinic with progressive weakness and decreased sensation of the right lower extremity, and sensory abnormality exhibiting stocking distribution. In electromyolography (EMG), demyelinating sensory motor polyneuropathy was determined. Ten years later, he was admitted again due to progression of weakness in the four extremities. Pyramidal and cerebellar signs, and loss of deep sensation were included in the clinical picture. An EMG investigation revealed severe segmental demyelination. Hyperintense lesions were determined in the periventricular deep white matter and the corpus callosum using cranial magnetic resonance imaging (MRI). This patient was considered to be exhibiting a course of demyelinating disease in which PNS and CNS were involved. In order to eliminate other reasons for peripheral neuropathy (PN), a sural nerve biopsy was carried out. The expression of several extracellular matrix (ECM) proteins (fibronectin, laminin, collagen type-IV), their respective receptors (integrin a5 and b4 subunits), intermediate filaments (vimentin) and S-100 protein was investigated by means of immunohistochemical methods. In addition, peripheral nerve tissue samples were evaluated ultrastructurally. Immunohistochemical stainings revealed an increase in the expression of ECM molecules, such as fibronectin, laminin and collagen type-IV, and their respective receptors, a5 and b4. This alteration might be a result of a Schwann-axon relationship. Vimentin expression was found to have changed in the Schwann cells and S-100 immunoreactivity decreased in the region near the Schwann-axon interface. Our ultrastructral findings showed myelin fragmentation, axon vacuolisation and degeneration.

Key Words: multiple sclerosis, sural nerve, fibronectin, laminin, integrin a5 and b4 subunits

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