38. **MULTIFOCAL ACQUIRED DEMYELINATING SENSORY AND MOTOR NEUROPATHY (MADSAM) WITH CRANIAL NERVE INVOLVEMENT. CASE REPORT**

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https://www.youtube.com/watch?v=vlsNiqV1-28&t=10174s

BACKGROUND: The multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) is a rare adult onset subvariant of the chronic inflammatory demyelinating polyneuropathy (CIDP). The latter affects 1-9 cases per million adults and may pose a diagnostic challenge (antibody and electrophysiological overlap). The hallmark of the disease spectrum is an immune mediated structural myelin breakdown. Urgent differentiation of MADSAM from the other variants of CIDP is indicated due to its distinct pathogenesis, management, and long-term therapy response to immunomodulators. The disease presents with progressive asymmetrical motor and sensory deficits initially located typically in one limb. Rarely the disease manifests with cranial nerve involvement. The prognosis of patients suffering from MADSAM is reliant on rapid diagnosis and therapeutic response, but neurological deficits reside. Due to its rarity and diagnostic challenge, misdiagnosis is common. **THE CASE**: A 84 years old Caucasian male diagnosed with multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) presented to our department with progressive neurological deficits. His neurological examination and history revealed paresthesia, hypesthesia, diminished vibration sense (pallhypesthesia), distally pronounced paresis in the upper extremities with a wrist drop on the left hand. Gait examination exposed coordination deficits. The history of the patient and the initial presentation of the disease in 1998 exemplifies the diagnostic challenge of MADSAM, due its mimicry of other diseases, like multifocal motor neuropathy (MMN). The initial symptoms were asymmetrical motor and sensory deficit starting at the upper extremities. Before the patient presented to our facility, he was diagnosed with neuroborreliosis and treated accordingly. The correct diagnosis was set at our department according to clinical presentation, nerve conduction velocity parameters and conduction block during electroneurography, typical changes in nerve ultrasound suggesting an inflammatory neuropathy and fasciculations of the gastrocnemius muscle. After an ineffective steroid therapy and long standing intravenous immunoglobulins the patient is now on a therapeutic scheme of 1000mg Rituximab (CD20 Ab) every 6 months and 1000mg Mycophenolatmofetil. The patient subjectively reports improvement of his status and slower progression of the disease since the Rituximab paradigm. Furthermore, an evident atrophy of the orbicularis oculi muscle was now noted during follow up examination, which highlights facial nerve involvement. **CONCLUSION**: This case exemplifies the challenge of diagnosing the multifocal acquired sensory and motor neuropathy, but prompt diagnosis and therapeutic intervention is associated with a better prognosis and slower progression of the disease in patients suffering from MADSAM.

Key words: Polyneuropathies; Electrophysiology; Facial Nerve (Source: MeSH-NLM).