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