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Letter to the Editor

Long-term observations in asymmetric immune-mediated neuropathy with vagus hypertrophy using ultrasound of the nerves

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1. Background

The clinical presentation of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) regularly consists of progressive or relapsing–remitting symmetric distal sensory disturbances in hands and feet as well as proximal flaccid paraparesis [1]. Diagnosis is made by nerve conduction studies (NCS) and increased cerebrospinal fluid protein without significant pleocytosis. However, variants of CIDP exist, e.g., the multifocal acquired demyelinating sensory and motor neuropathy (MADSAM). Treatment options for all CIDP-variants include systemic steroids, immunoglobulins (IVIg) and immunosuppressants [2]. Early diagnosis may be difficult.

We recently reported a patient with asymmetric immune-mediated neuropathy with prominent dysphonia/dysphagia and flaccid tetraparesis [3]. We described considerable swelling of the peripheral nerves and the vagus, detected by ultrasound.

The role of ultrasound as diagnostic tool particularly in immune-mediated polyneuropathies is gaining importance [4]. It was described as an observation method for therapy response, however the time course and degree of nerve changes remain unclear so far [4,5].

Here, we describe the follow-up values over twelve months.

1.1. Clinical course at onset, after six and twelve months

At onset, the patient complained about hoarseness and dysphagia since several weeks. On demand, the patient remembered slight asymmetric sensorimotor deficits in hands and thighs since one year. The examination revealed left-sided vocal cord palsy, flaccid tetraparesis (MRC-Score 50) and reduced/absent deep tendon reflexes. Further, we found sensory deficits in both hands and feet. According to the NCS (e.g., temporal dispersion, conduction blocks) mentioned below, the results of a nerve biopsy and the cerebrospinal fluid, and after exclusion of possible mimics (e.g., inherited neuropathies/neurofibromatosis) we diagnosed a CIDP (possibly MADSAM due to asymmetry and cranial nerve involvement) and initiated a therapy with IVIg 0.4 g/kg bodyweight for 5 days, followed by weekly infusions.

Six months later, hoarseness and tetraparesis were improved (MRC-Score 56). IVIg was reduced to once/second week. After treatment over twelve months, hoarseness, dysphagia and paresis completely resolved (MRC-Score 60), and the only gait imbalance persisted.

1.2. NCS at onset, after six and twelve months

Table 1 gives an overview of NCS, which fulfilled the EFNS-guideline criteria for CIDP. After twelve months, the NCS results were heterogeneously changed, ranging from improvement to increased (secondary) axonal loss. E.g., the extent of axonal damage (as assessed by CMAP amplitude reduction) increased in the peroneal nerve. CV reduction as well as the incomplete conduction block in the median, the ulnar and the tibial nerves improved slightly.

Table 1
Nerve conduction and ultrasound follow-up values.

Ultrasound CSA in mm^2	Reference boundary values [6,7]	Onset	Six months	Twelve months	Electrophysiology	Onset	Twelve months
MN UA right/left	**10 mm^2**	**102/110**	102/110	**62/44**	MN dmL/CV right	4.4 ms/27 m/s	4.0 ms/31 m/s
MN FA r/l	10 mm^2	98/**79**	85/68	99/**36**	MN CMAP right dist/prox	**8.2/3.7 mV**	**10/5.8 mV**
ICSAV right/left		**10.1**	NA	**5.7**	F-wave response	55.7 ms	68.9 ms
UN UA right	**9 mm^2**	**66**	NA	**30**	UN dmL/CV left	**4.8 ms/30 m/s**	**3.5 ms/35 m/s**
UN FA right	8 mm^2	34	NA	27	UN CMAP right dist/prox	4.2/2.7 mV	6.2/3.2 mV
ICSAV right		8.3	NA	7.1	MN SNAP/CV	10 μV/48 m/s	4 μV/56 m/s
C6 diameter in mm	3.8 mm	4.8	4.2	4.2	PN dmL/CV right	3.9 ms/44 m/s	5.6 ms/45 m/s
SUR right	2.5 mm^2	6	NA	5	PN CMAP dist/prox	1.5/1.7 mV	0.3/0.3 mV
TN P right	**30 mm^2**	**64**	64	**40**	TN dml/CV left	7.8 ms/37 m/s	6.0 ms/32 m/s
TN A left	10.5 mm^2	21	36	22	TN CMAP dist/prox	**11.7/4.3 mV**	**13.0/8.2 mV**
PN left	**11 mm^2**	**19**	17	**13**	F-wave response	66.2 ms	72.6 ms
Vagus right	**2.5 mm^2**	**9**	**6**	**4**	SN SNAP/CV	5 μV/48 m/s	4 μV/50 m/s

Abbreviations: a = ankle; C6 = cervical root 6; CMAP = compound muscle action potential amplitude; CSA = cross sectional area; CV = conduction velocity; dmL = distal motor latency; dist = distal; FA = forearm; ICSAV = intra-nerve CSA variability; l = left; MN = median nerve; mm = millimeter; mm^2 = square millimeter; ms = millisecond; mV = millivolt; m/s = meter per second; UA = upper arm; UN = ulnar nerve; P = poplitea; PN = peroneal nerve; prox = proximal; r = right; SNAP = sensory nerve action potential; sur = sural nerve; TN = tibial nerve. Remarkable differences are in bold.

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1.3. Ultrasound at onset, after six and twelve months

Table 1 gives an overview of the ultrasound values at different time points compared to boundary values [6,7]. All nerves were scanned along their courses and the cross-sectional area (CSA) was measured at predefined measurement points including maximum/minimum values. Additionally, we measured the diameter of the cervical root 5/6. Nerves and cervical roots revealed considerable

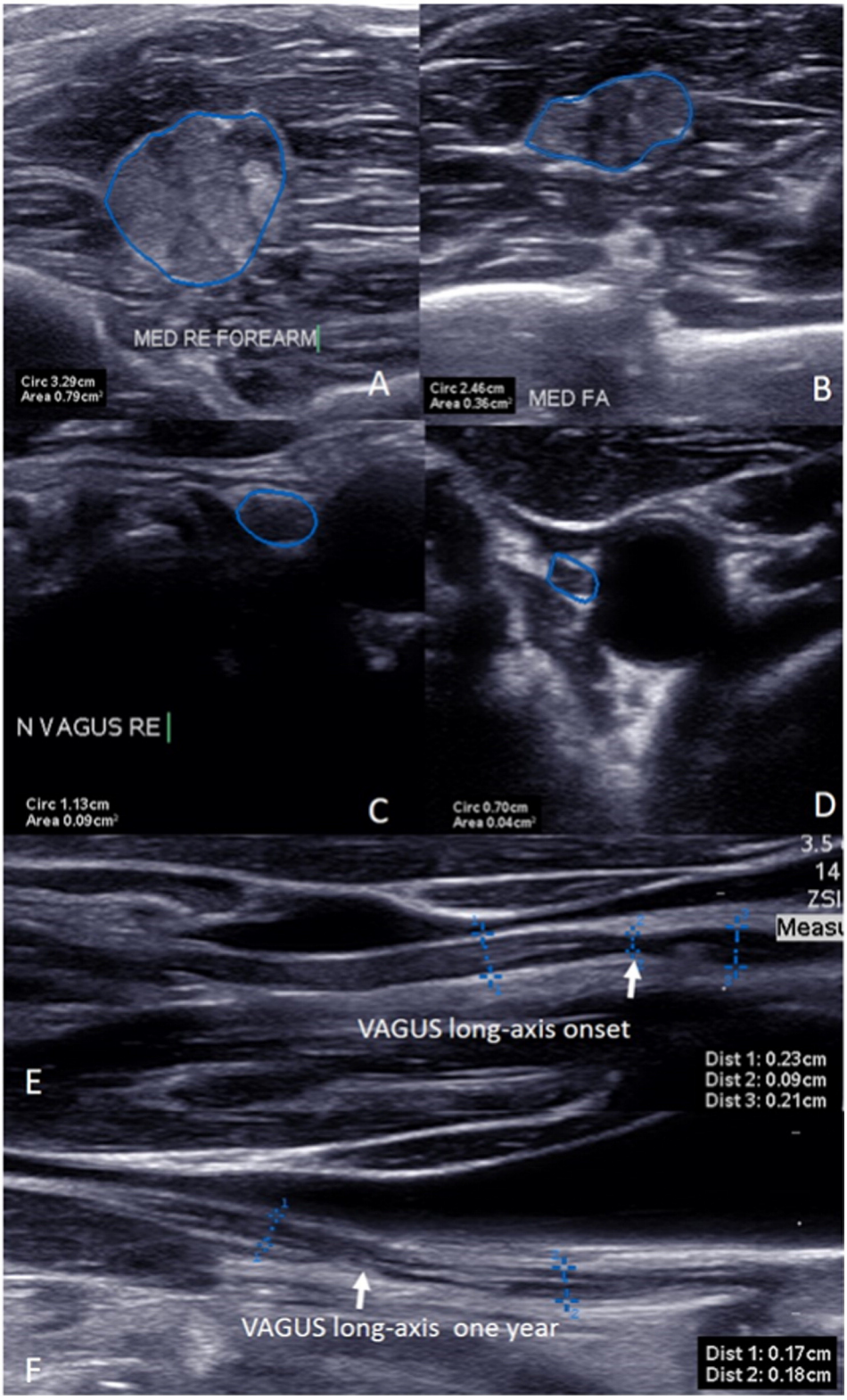

Fig. 1. Short-axis of the median nerve in the right forearm at onset (A, CSA 79 mm^2) and after twelve months (C, 36 mm^2). Echogenicity at onset is remarkably increased. C shows the short-axis vagus nerve at onset (9 mm^2) and after twelve months (D, 4 mm^2) next to carotid artery. E and F show the long-axis of the vagus at onset (E, diameter 2.1 and 2.3 mm with caliber reduction in one part to 0.9 mm, the echointensity seems to be reduced) and after twelve months (F, diameter 1.7 and 1.8 mm, the echointensity seems to be normalized).

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