Journal of the Neurological Sciences 348 (2015) 211–215

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Bochum ultrasound score allows distinction of chronic inflammatory from multifocal acquired demyelinating polyneuropathies

A. Kerasnoudis *[,1], K. Pitarokoili [2], R. Gold [3], M.-S. Yoon [4]

Department of Neurology, St. Josef Hospital, Ruhr-University of Bochum, Germany

ARTICLE INFO

Article history:
Received 1 October 2014
Received in revised form 9 November 2014
Accepted 8 December 2014
Available online 11 December 2014

Keywords:
Bochum ultrasound score
Chronic inflammatory demyelinating polyneuropathy
Multifocal motor neuropathy
Multifocal acquired demyelinating sensory and motor neuropathy
Nerve ultrasound
Polyneuropathies

ABSTRACT

Objective: The aim of this observational study was to evaluate the applicability of a recently introduced ultrasound score (Bochum ultrasound score; BUS) in distinguishing the chronic inflammatory demyelinating polyneuropathy (CIDP) from the multifocal motor neuropathy (MMN) or the multifocal acquired demyelinating sensory and motor neuropathy (MADSAM).
Methods: The BUS underwent prospective evaluation of its applicability in a group of 13 patients (mean age 47.2, SD ±13.7, 9 women), who were referred to our department between January 2012 and August 2013 with the clinical picture of a chronic symmetrical or asymmetrical sensory/sensorimotor neuropathy.
Results: The cut-off value of ≥2 points in the "Bochum ultrasound score" showed a sensitivity of 80% and specificity of 87.5% (PPV = 80%, NPV = 87.5%) in distinguishing CIDP from MMN or MADSAM.
Conclusions: The BUS seems to allow a reliable distinction of CIDP from multifocal acquired demyelinating polyneuropathies causing predominantly motor nerve dysfunction, such as MMN or MADSAM. Our ultrasound findings indicate a stronger relationship of MADSAM to MMN, than to CIDP.

1. Introduction

Multifocal motor neuropathy (MMN) is an intriguing peripheral nerve disease with a prevalence of 1 to 2 per 100,000 adults mainly affecting men and young adults [19,20]. Several diagnostic criteria have been proposed, summarizing the asymmetric slowly progressive weakness, with a striking predilection for the upper extremities, whereas sensory fibres and upper motor neuron signs fail in the disease course [7]. Although the detection of conduction block remains the electrophysiological hallmark of the disease, it is important to recognize, that it may be not possible to demonstrate this finding even after careful studies. The reason is, that these blocks may be activity-dependent, or the site of pathology may be very proximal in the brachial plexus or nerve root level [21,25]

Typical chronic inflammatory demyelinating polyneuropathy (CIDP) arises between the ages of 30 and 60 years and is characterised by a progressive, symmetric, proximal and distal muscle weakness, paresthesis, sensory dysfunction and impaired balance, which may evolve slowly over at least 2 months [16,29]. Lewis et al. reported in 1982 five patients with chronic demyelinating neuropathy with clinical signs of mononeuritis multiplex and electrophysiological evidence of multifocal persistent conduction block [18]. In addition, Saperstein et al. described 11 patients with multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) and compared them with 16 MMN patients, concluding that MADSAM neuropathy more closely resembles CIDP and probably represents an asymmetrical and atypical variant [26]. The nosological position of this neuropathy in relation to MMN and CIDP is still debated. Various reports followed, mainly considering MADSAM as a variant of CIDP [23,24,30], or an intermediate link between CIDP and MMN [3,23], or a distinct clinical entity [23,30].

The diagnostic challenge of distinguishing these three immune-mediated polyneuropathies remains high, as in the case of CIDP or MADSAM corticosteroids exert short term or long term clinical improvement in about two-thirds of patients [17], while MMN does not improve with steroids in most of the patients [6,8,32]. Rash distinction between these three entities consecutively leading to an early and accurate immune therapy might be of great help for assessment of clinical outcome and prognosis.

* Corresponding author at: Dept. of Neurology, Ruhr University, St. Josef-Hospital, Gudrunstr. 56, 44791 Bochum, Germany. Tel.: +49 17662923235; fax: +49 2345093024.
E-mail addresses: antonis.kerasnoudis@gmail.com (A. Kerasnoudis), kalliapit@yahoo.gr (K. Pitarokoili), Ralf.Gold@ruhr-uni-bochum.de (R. Gold), min-suk.yoon@uni-due.de (M.-S. Yoon).

[1] Antonios Kerasnoudis acquired, analyzed and interpreted the data of this study.
[2] K.Pitarokoili acquired and analyzed the data of this study.
[3] R. Gold made the critical revision of this study.
[4] M.-S. Yoon acquired, analysed, interpreted the data and made the critical revision of this study.

http://dx.doi.org/10.1016/j.jns.2014.12.010
0022-510X/

While recent ultrasound studies reported significant differences of the pathological findings among axonal and demyelinating polyneuropathies [5], the recent introduction of a new ultrasound score (Bochum ultrasound score; BUS) allowed the differentiation of subacute CIDP from acute demyelinating polyneuropathy (AIDP) [9]. The aim of our study was to evaluate prospectively the applicability of the Bochum ultrasound score in the distinction of CIDP from MMN or MADSAM.

2. Methods

2.1. Patients

The local university ethics committee approved our study protocol and all CIDP, MMN and MADSAM patients signed informed consent. For the diagnosis of definite CIDP or MMN we used the diagnostic criteria proposed from the Joint Task Force of the European Federation of Neurological Societies and the Peripheral Nerve Society [7,31]. On the other hand, MADSAM patients were defined as a clinical presentation of asymmetrical sensory or sensorimotor neuropathy with definite persistent conduction blocks [22]. Extensive clinical and laboratory evaluation excluded other causes of neuropathy.

2.2. Study protocol

For the prospective evaluation of the Bochum ultrasound score in distinguishing CIDP from MMN or MADSAM, 13 patients, who referred to our department between January 2012 and August 2013 with clinical presentation of symmetrical or asymmetrical sensory/sensorimotor neuropathy underwent a double blinded electrophysiological and ultrasound evaluation.

2.3. Ultrasound examination

Ultrasonography was performed blinded for the clinical and electrophysiological findings from a board certified neurologist (A.K.) for neuromuscular ultrasound. All ultrasound studies have been performed with the use of an Aplio® XG ultrasound system (Toshiba Medicals, Tochigi, Japan). For the superficial nerves of the body (ulnar, radial, and sural) an 18-MHz linear array transducer was used. Each nerve was identified, and care was taken to adjust the transducer so it was perpendicular to the nerve and the smallest cross-sectional image was obtained. Cross sectional area measurements in each of the nerves were performed at the inner border of the thin hyperechoic epineural rim by the continuous tracing technique and the average values were calculated after serially measuring three times. The intrarater reliability was determined with the help of the dependability coefficient (phi) after measuring the CSA of the ulnar nerve in Guyon's canal on a single healthy control in 5 consecutive days.

In view of our recent report [9], the anatomical sites summarised under the "Bochum ultrasound score" included the cross sectional area of: a) the ulnar nerve in Guyon's canal, b) the ulnar nerve in upper arm, c) the radial nerve in spiral groove and d) the sural nerve between the lateral and medial head of the gastrocnemius muscle (Table 1, Fig. 1). The scoring system included two simple rules 1) the patient received 1 point, for each of the above anatomic sites, where he showed a pathological cross sectional area enlargement, compared with reference values of our lab [14], 2) if the patient showed on both sides of the body a pathological cross sectional area nerve enlargement of the concrete nerve, he also received only 1 point. Considering the above, each patient could receive a minimum sum score of 0 point and a maximum sum score of 4 points. Following the previous report in the literature [9] we used as a cut-off value for differentiating a CIDP from MMN or MADSAM a sum score of ≥2 points.

Table 1
Overview of the anatomic sites and scoring system of the Bochum Ultrasound score.

Anatomic sites	Points
CSA of the ulnar nerve in Guyon's canal	1
CSA of the ulnar nerve in the upper-arm	1
CSA of the radial nerve in spiral groove	1
CSA of the sural nerve between the gastrocnemius muscle	1
Sum Score	**4**

The scoring system included two rules 1) the patient received 1 point, for each of the above anatomic sites, where he showed a pathological cross sectional area enlargement, compared with the reference values of our lab [14], 2) if the patient showed on both sides of the body a pathological cross sectional area nerve enlargement of the concrete nerve, he also received only 1 point. Considering the above, each patient could receive a minimum sum score of 0 point and a maximum sum score of 4 points. CSA = cross sectional area.

2.4. Nerve conduction studies

All electrophysiological studies were performed blinded for the clinical and ultrasound findings from a board certified neurologist (M.-S. Y.) with the use of a Medtronic 4 canal electromyography device (Medtronic, Meerbusch, Germany). All testing was done while maintaining the skin temperature at 36 °C. Motor and sensory studies were performed in the median and ulnar nerve. In addition, motor studies were performed in the fibular and tibial nerve and sensory studies in the sural nerve. Sural potentials were recorded from antidromic stimulation at the lower lateral third of the mid-calf, the recording electrode being located below the lateral malleolus. Averaging of at least ten responses was performed for all sensory studies to improve signal to noise ratio. We used for the detection of conduction block the consensus criteria proposed from the American Association of Electrodiagnostic Medicine [1].

2.5. Statistics

For each type of immune neuropathy studied (CIDP, MMN, and MADSAM) sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) were calculated. Taking as example the CIDP, sensitivity was defined as: number of true positive patients for CIDP / number of false negative patients for CIDP + number of true positive patients for CIDP, while specificity was defined as: number of true negative patients for CIDP / number of false positive patients for CIDP + number of true negative patients for CIDP. On the other hand, positive predictive value (PPV) was defined as: number of true positive patients for CIDP / number of false positive patients for CIDP + number of true positive patients for CIDP, while negative predictive value (NPV) was defined as: number of true negative patients for CIDP / number of false negative patients for CIDP + number of true negative patients for CIDP.

3. Results

The BUS underwent prospective evaluation in a group of 13 patients (mean age 47.2, SD ± 13.7, 9 women), who referred to our department with clinical presentation of symmetrical or asymmetrical sensory/sensorimotor neuropathy. All patients underwent a double blinded electrophysiological and ultrasound evaluation a mean of 2.8 years (SD ± 1.5, min 3 months, max 4 years) after disease onset. The final diagnosis in each of the 13 cases was based on clinical and electrophysiological follow up. Hence, 5 patients fulfilled the diagnostic criteria for CIDP [31], 6 for MMN [7] and 2 patients for MADSAM [22]. The results of the nerve conduction and ultrasound studies of the 13 patients are provided in Tables 2 and 3. The intrarater reliability of the ultrasound examiner was adequate (phi > 0.946).

The cut-off value of ≥2 points in the "Bochum ultrasound score" showed a sensitivity of 80% and specificity of 87.5% (PPV = 80%, NPV = 87.5%) in distinguishing CIDP from MMN or MADSAM (Table 4).