A CASE REPORT: APPLICATION OF THE UPSS SCORING SYSTEM IN THE DIAGNOSIS OF CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

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ABSTRACT

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is an acquired immune-mediated peripheral neuropathy characterized by progressive motor weakness and sensory deficits affecting both upper and lower limbs. The pathophysiology involves recurrent demyelination and remyelination of peripheral nerves, leading to segmental conduction block and impaired nerve signal transmission. Diagnosis is often challenging due to clinical and electrophysiological overlap with other neuropathies such as Guillain–Barré syndrome (GBS)/Acute Inflammatory Demyelinating Polyneuropathy (AIDP) and Chronic Immune Sensory Polyradiculopathy (CISP). The Ultrasound Pattern Sum Score (UPSS), particularly the UPSS-A component, has been shown to be a useful tool in the differential diagnosis of CIDP.

We report a case of a 20-year-old male with no prior medical history, admitted for progressive numbness in both lower limbs and upper limbs, reduced deep tendon reflexes, and diminished vibration sense. Electromyography (EMG) showed reduced motor and sensory conduction velocities in multiple peripheral nerves bilaterally. Peripheral nerve ultrasound demonstrated hypoechoic, multifocal nerve enlargement involving various fascicles and segments. UPSS scores were as follows: UPSS-A: 14; UPSS-B:1; UPSS-C: 1; total score =16. The final diagnosis was CIDP.

Keywords: UPSS, Chronic inflammatory demyelinating polyneuropathy, CIDP, peripheral nerve ultrasound.

I. CASE REPORT

A 20-year-old male patient previously in good health was admitted with complaints of numbness in both lower limbs, progressing proximally to the upper limbs in a glove-and-stocking distribution. Subsequently, he developed gradually worsening lower limb weakness, leading to difficulty in ambulation. Bowel and bladder functions remained intact, and there were no associated cranial nerve deficits.

Neurological examination revealed distal muscle weakness in both upper limbs (Medical Research Council scale [MRC] grade 4/5) and lower limbs (MRC grade 3/5), decreased deep tendon reflexes, and diminished vibration sense.

Blood biochemistry test, complete blood count, and cerebrospinal fluid (CSF) analysis were within normal limits. Magnetic Resonance Imaging (MRI) of the brain and cervical spine showed no abnormalities.

Electromyography (EMG) demonstrated reduced motor and sensory conduction velocities in the ulnar, median, fibular, and sural nerves, as well as reduced motor conduction in the tibial nerve. Peripheral nerve ultrasound revealed hypoechoic and multifocally enlarged nerve fascicles, with variations in size along different segments of the nerves. The patient was assessed using the Ultrasound Pattern Sum Score (UPSS) with the following results: UPSS-A = 14 points, UPSS-B = 1 point, and UPSS-C = 1 point, giving a total score of 16.

The final diagnosis was Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP).

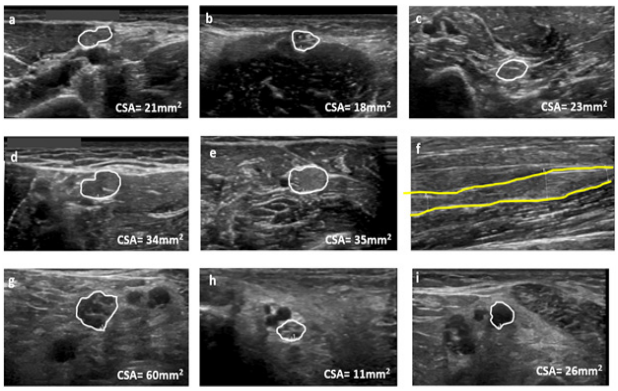

Figure 1: Ultrasonographic image of the right peripheral nerves at median forearm (a), elbow (b), mid-arm (c); ulnar forearm (d), mid-arm (e); tibial knee (g), ankle (h); and fibular knee (i) of one of the CIDP patients. Total UPSS - A score: (2 + 2 + 2 + 2 + 2 + 2 + 0 + 2) = 14. (f): uneven hypertrophy of the right median nerve in the mid-arm.

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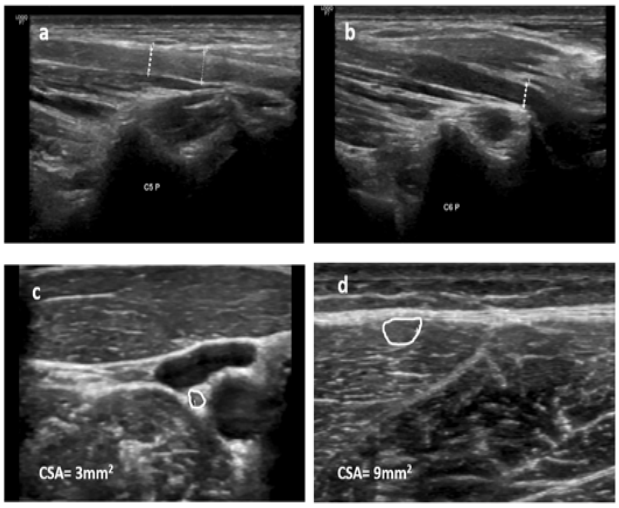

Figure 2: Ultrasonographic image of the right peripheral nerves at the C5 root (a) with a diameter of 2.5mm, C6 (b) with a diameter of 4.7mm, the vagus nerve (c) and the sural nerve (d). UPSS - B score = 0 + 1 + 0 = 1. UPSS - C score = 1.

II. DISCUSSION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is the most common treatable chronic immune-mediated neuropathy, characterized by progressive or relapsing sensorimotor dysfunction resulting from immune-mediated demyelination of peripheral nerves [1,2]. Early and accurate diagnosis is crucial because CIDP is potentially reversible with immunomodulatory therapies [2]. However, the diagnosis remains challenging due to the heterogeneity of clinical presentations and overlap with other peripheral neuropathies, particularly Guillain–Barré syndrome (GBS), hereditary neuropathies such as Charcot–Marie–Tooth disease (CMT), and metabolic neuropathies including diabetic neuropathy [2,3].

In the present case, the patient exhibited gradually progressive sensory disturbances and distal limb weakness over the course of one month, accompanied by reduced deep tendon reflexes and electrophysiological evidence of demyelination. These findings are consistent with the classical clinical phenotype of CIDP. Nevertheless, distinguishing CIDP from acute inflammatory demyelinating polyneuropathy (AIDP) can be difficult in the early stages because of overlapping clinical and electrophysiological features [2].

Peripheral nerve ultrasound has emerged as a valuable adjunctive diagnostic tool in the evaluation of immune-mediated neuropathies [4,5]. Previous studies have demonstrated that patients with CIDP typically exhibit multifocal and asymmetric enlargement of peripheral nerves, reflecting inflammatory edema and repeated cycles of demyelination and remyelination [4,6]. In contrast, hereditary neuropathies such as CMT often show diffuse and homogeneous nerve enlargement, whereas GBS tends to demonstrate less pronounced or more proximally restricted changes [6]. Therefore, the pattern of nerve enlargement assessed by ultrasound may provide important diagnostic clues.

To standardize the interpretation of nerve ultrasound findings, the Ultrasound Pattern Sum Score (UPSS) was introduced as a quantitative scoring system that evaluates nerve enlargement across predefined anatomical regions [7]. The UPSS incorporates three components: enlargement of peripheral nerves in the upper limbs (UPSS-A), lower limbs (UPSS-B), and cervical nerve roots or vagus nerve (UPSS-C) [7]. Higher total UPSS scores are strongly associated with CIDP, whereas lower scores are more frequently observed in other neuropathies [7,9]. Previous studies have reported that a UPSS above certain thresholds demonstrates good sensitivity and specificity for distinguishing CIDP from other demyelinating neuropathies [8,9].

Component	Measurement Sites and Reference Values	CSA Score >150%	CSA Score 100-150%	Maximum Score
UPS-A	8 sites: - Median nerve at upper arm (8.3 mm^2) - Median nerve at elbow (8.3 mm^2) - Median nerve at forearm (6.4 mm^2) - Ulnar nerve at upper arm (5.9 mm^2) - Ulnar nerve at forearm (5.2 mm^2) - Tibial nerve at popliteal fossa (25.9 mm^2) - Tibial nerve at medial malleolus (12.7 mm^2) - Peroneal nerve at popliteal fossa (9.2 mm^2)	2	1	16
UPS-B	3 sites: - C5 root (5.6 mm^2) - C6 root (8.8 mm^2) - Vagus nerve (2.2 mm^2)	2	1	3
UPS-C	1 site: - Sural nerve (3.5 mm^2)	1	1	1
Total UPSS				0-20

Figure 3: The ultrasonic pattern sum score UPSS [3]

In our patient, peripheral nerve ultrasound revealed hypoechoic and multifocal enlargement affecting multiple nerve segments and fascicles. The calculated UPSS values (UPSS-A = 14, UPSS-B = 1, UPSS-C = 1) resulted in a total score of 16, which strongly supported the diagnosis of CIDP. Notably, the prominent enlargement of upper limb nerves reflected by the high UPSS-A score aligns with previously described ultrasound patterns of CIDP [4,8].

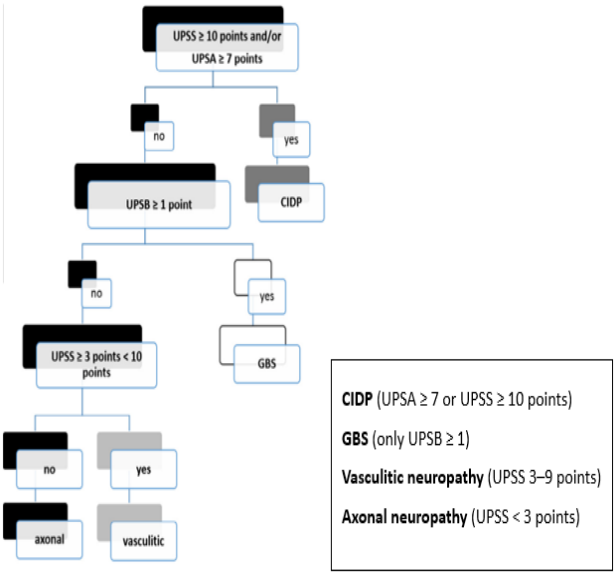

Figure 4: Suggestion of an algorithm how to use the UPSS in daily routine.[3]

Abbreviations: axonal = axonal non immune-mediated neuropathies; CIDP = chronic inflammatory demyelinating; polyradiculoneuropathy; GBS = Guillain–Barré syndrome; UPSS = ultrasound pattern sum score.

The present case highlights the clinical utility of the UPSS scoring system as a complementary diagnostic tool in suspected CIDP. Compared with electrophysiological testing alone, the integration of nerve ultrasound allows visualization of structural nerve changes and provides additional information regarding the distribution and pattern of nerve involvement [4,5]. This is particularly valuable in cases where conventional diagnostic tests such as cerebrospinal fluid analysis or MRI fail to demonstrate abnormalities.

Moreover, the non-invasive, rapid, and relatively accessible nature of nerve ultrasound makes it an attractive modality in routine clinical practice [4]. The use of standardized scoring systems such as UPSS further improves reproducibility and facilitates objective comparison across patients and institutions [7,8,10]. As the role of neuromuscular ultrasound continues to expand, it may become an important component of the diagnostic algorithm for immune-mediated neuropathies [5].

Nevertheless, several limitations should be acknowledged. First, nerve ultrasound findings may vary depending on the stage and subtype of CIDP, and overlap with other neuropathies may still occur [5,6]. Second, the interpretation of ultrasound results is operator-dependent and requires adequate expertise. Finally, as this report describes a single case, further studies with larger patient cohorts are needed to validate the diagnostic performance of the UPSS scoring system in different clinical settings [8,9].

In conclusion, this case demonstrates that the integration of peripheral nerve ultrasound and the UPSS scoring system can provide valuable diagnostic support in suspected CIDP. The characteristic pattern of multifocal nerve enlargement and a high UPSS score may help differentiate CIDP from other demyelinating neuropathies, particularly in cases with inconclusive conventional investigations [4,7]. Incorporating ultrasound-based scoring systems into the diagnostic workflow may enhance early recognition and facilitate timely treatment of this potentially reversible neuropathy [2].

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