Neuromuscular Ultrasound Clinical Case Study

Ultrasound diagnostics in CIDP

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Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare autoimmune disorder of the peripheral nervous system, characterized by immune-mediated inflammation primarily affecting the myelin sheath. CIDP predominantly affects males with a peak incidence of 40 to 60 years. CIDP is a progressive or relapse-remitting neuropathy developing over more than 8 weeks. The typical form is characterized by symmetric proximal and distal muscle weakness with areflexia of the four limbs and sensory dysfunction with a predominance of large fibers involvement (paresthesia's, hypoesthesia, ataxia), leading to impaired balance and walking. Many variants of CIDP exist, such as pure sensory, pure motor, or asymmetric CIDP, representing almost half the patients, which makes diagnosis often complex. Despite CIDP being a rare disease with a prevalence around 0.8 to 8.9 per 100,000 people, this condition has been drawing growing interest within the field of neurology because treatment exist. Therefore, early diagnosis is crucial for therapeutic management and to improve prognosis.

The role of ultrasound in CIDP diagnosis

Electrodiagnostic studies (EDX) are mandatory for CIDP diagnosis by showing nerve conduction abnormalities suggestive of demyelination. Nonetheless, EDX sensitivity and specificity are not perfect, and other paraclinical investigations (i.e. supportive criteria) should be considered when diagnosis is uncertain after clinical and EDX assessment[1]. Ultrasound imaging of nerves has recently emerged as a valuable addition to the diagnostic toolkit of peripheral neuropathies, with the obvious advantages of its easy applicability and non-invasiveness. Nerve ultrasound can help differentiate demyelinating polyneuropathies from axonal neuropathies, by demonstrating multifocal or generalized enlargement of the nerve, in hereditary and inflammatory neuropathies[2]. In typical CIDP, nerve ultrasound reveals diffuse or segmental nerve hypertrophy, modification of fascicle size and morphology and alteration of nerve echogenicity. Proximal median nerve and brachial plexus are the most commonly affected segments. Cross sectional area (CSA) reference values for peripheral nerves and brachial plexus have been reported in various studies in the literature, and can be easily used[3-4]. A short scanning protocol has been recently published for CIDP diagnosis, and has proven its efficiency to diagnose CIDP in patients with a clinical suspicion but without demyelination criteria on EDX[5]. The combination of EDX and nerve ultrasound allow 20-30%[3] additional CIDP diagnosis. Finally, nerve ultrasound has been shown to improve diagnostic accuracy in a significant way, making it easier to identify patients with treatable inflammatory neuropathies. It also provides a non-invasive way to monitor changes over time, even if it is still not clear how nerve size and morphology varies with time and therapeutic response.

Case study

A 58-year-old patient with no previous medical history presented to the neurology department with sensory disturbances in both hands that had been evolving for 2 years. He had no motor complaints and no trouble walking. He experienced discomfort and slight disability in the course of his work (dressmaking). The clinical examination was normal, apart from abolition of tendon reflexes of the four limbs. Nerve ultrasound has been performed with Canon Aplio a450 machine using the 18 MHz high resolution transducer covering a bandwidth of 7 to 18 MHz. Striking abnormalities with enlargement of most of the nerves have been revealed, including sensory nerves (sural, fig. 1), motor nerves of four limbs (tibial nerves at popliteal fossa, fig. 2; median and ulnar nerves, figs. 3a-b and 4a-b) and cervical roots (fig. 5) and brachial plexus (fig. 6). There was also fascicular enlargement with great intranerve heterogeneity (coexistence of normal fascicules and enlarged fascicules, see figs. 2 and 3a). Enlargement was sometimes really focal, with a « fusiform » shape (fig. 3b). There was a clear distal-proximal gradient, with the most prominent abnormalities being at median and ulnar nerves at the arm (figs. 3-4). Echogenicity was variable, with some hypoechoic nerves (fig. 4a) and other with isoechoic component (figs. 4b). Brachial plexus in the supra-clavicular region were enlarged but clearly asymmetrical predominantly on the left one (fig. 6). Ultrasound was typical of a demyelinating neuropathy, and the intra-nerve and inter-nerve heterogeneity was evocative of an acquired demyelinating neuropathy. In combination with clinical and EDX assessment, CIDP was diagnosed. The patient was treated with 3 rounds of intravenous immunoglobulins with improvement of sensory complaints.

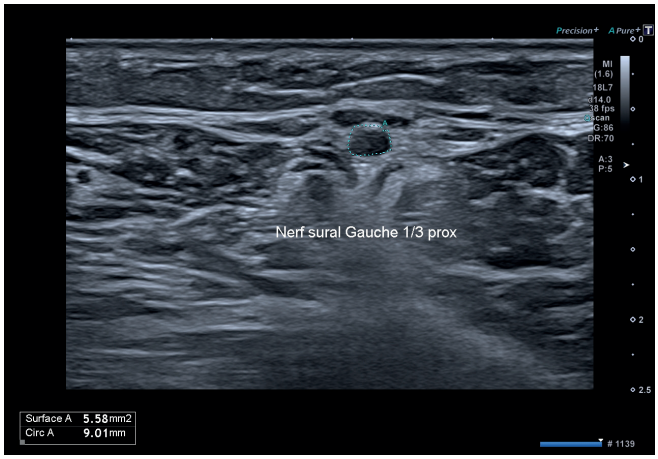

Figure 1 Left sural nerve enlargement, at the proximal part of the calf, CSA 5.58 mm^2.

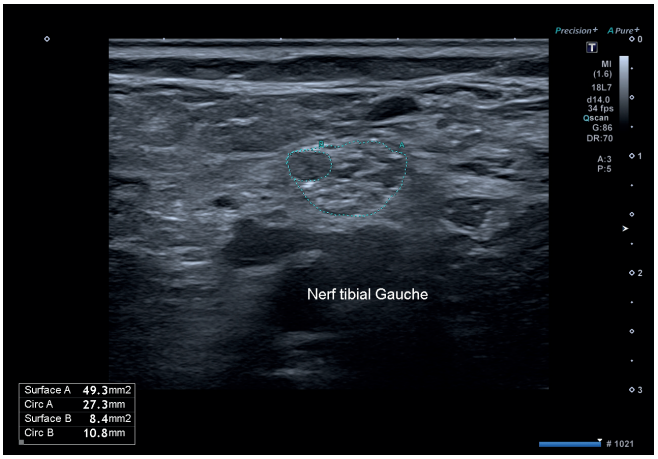

Figure 2 Left tibial nerve enlargement at popliteal fossa, CSA 49.3 mm^2 including enlarged fascicle measuring CSA of 8.4 mm^2.

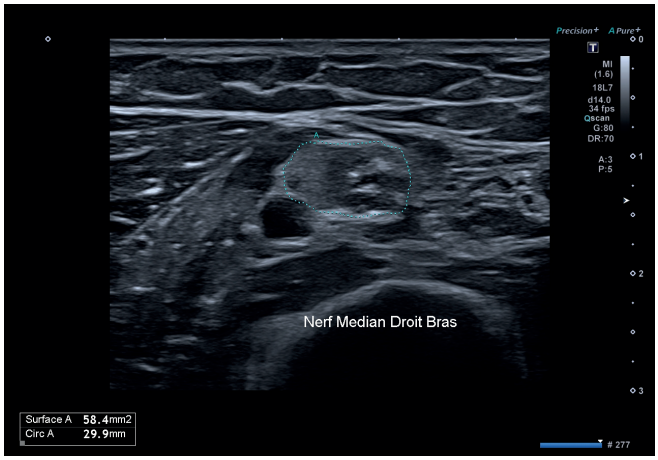

Figure 3a Right median nerve at upper arm, enlarged CSA (58.4 mm^2) and fascicles.

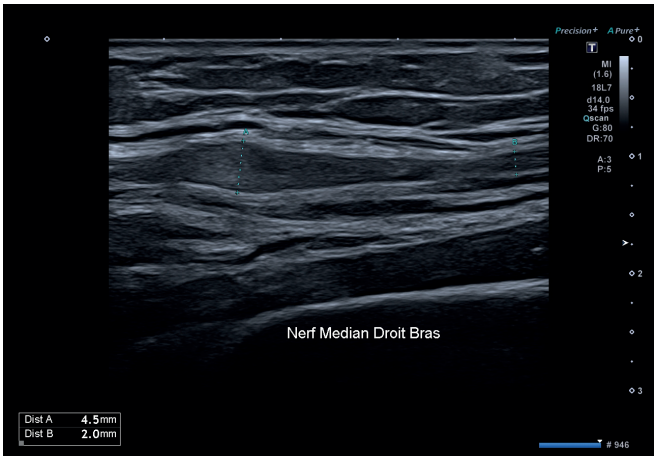

Figure 3b Right median nerve at upper arm, enlarged diameter with focal hypertrophy.

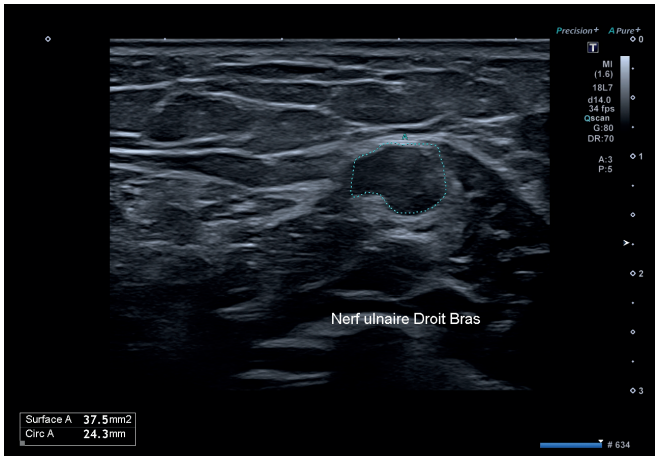

Figure 4a Right ulnar nerve at upper arm, enlarged CSA 37.5 mm^2 with hypoechoic nerve.

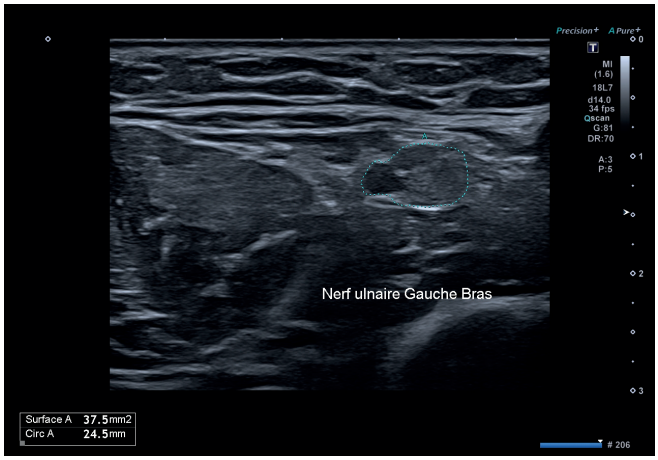

Figure 4b Left ulnar nerve at upper arm, enlarged CSA 37.5 mm^2.

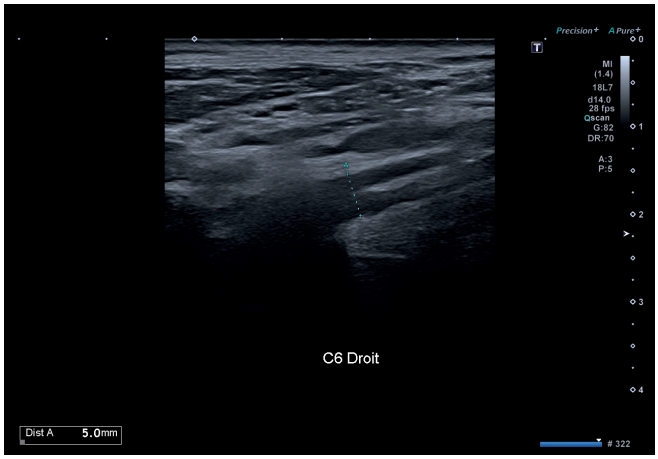

Figure 5 Cervical root C6, with an enlarged diameter (5 mm) at the exit of foramina.

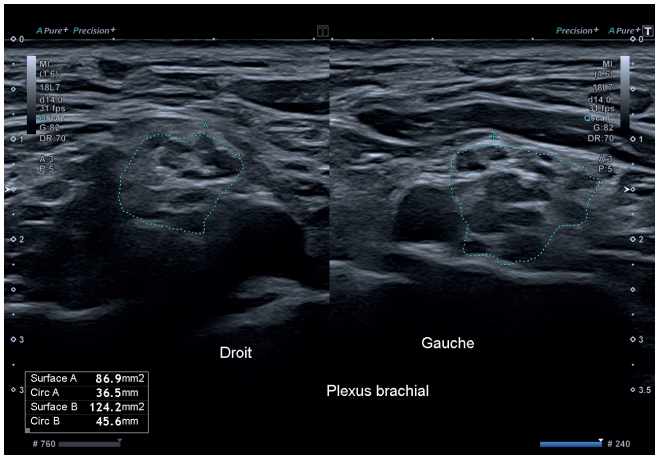

Figure 6 Right and left enlarged brachial plexus, predominant on the left side.

NERVE	CSA (cross-sectional area) in mm^2 and diameter (for cervical roots) in mm		
	Right	**Left**	**Normal values[4-5]**
Median			
Forearm	18.8	20.8	< 10 mm^2
Upper arm	58.4	33.4	< 13 mm^2
Ulnar			
Forearm	11	17.3	< 8.5 mm^2
Upper arm	37.5	37.5	< 9.5 mm^2
Cervical roots			
C5	3.2	4.1	< 2.9 mm
C6	5	4.2	< 3.8 mm
Tibial			
Popliteal fossa	53.8	49.3	< 33 mm^2
Sural	1.9	1.5	< 3.5 mm^2
Superficial fibular	2.1	2.4	< 3.5 mm^2

Figure 7 Report with measured CSA and diameter values of the patient

Conclusion

Overall, nerve ultrasound is a useful tool in the diagnosis of CIDP by showing multifocal or diffuse nerve enlargement and alteration of echogenicity. Progress is still needed on the value of ultrasound as a prognostic factor and as a means of monitoring response to treatment.

References

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