POSTER Session:
Malattie del Motoneurone
ID n. 470

Azienda Ospedaliero Universitaria Pisana
OSPEDALE DI RILIEVO NAZIONALE E DI ALTA SPECIALIZZAZIONE

NEUROMUSCULAR ULTRASOUNDS (NMUS) AS SUITABLE DIAGNOSTIC TOOL IN THE ASSESSMENT OF SEVERITY AND DISEASE PROGRESSION OF AMYOTROPHIC LATERAL SCLEROSIS.

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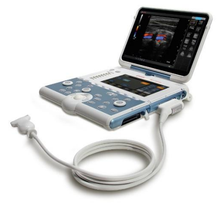

Introduction and Aims: Several methods have been proposed to diagnose and score disease severity in patients with Amyotrophic Lateral Sclerosis (pALS). Traditionally, neurophysiological tests have been employed. The recent development of neuromuscular ultrasound (US) has provided a further tool to supplement clinical and neurophysiological assessment of pALS. In the present study we evaluated sonographic changes of main muscles and nerve trunks in an attempt to improve diagnosis and assess disease severity.

Materials and Methods: 32 consecutive pALS (15 m and 17 f; mean age: 64.6 + 10.5 yrs) were enrolled. All investigations were performed using a Telemed Echo-wave II or a Esaote MyLabGamma device in conventional B-Mode. We evaluated the following muscles: sternocleidomastoid, diaphragm, biceps brachii/brachialis, forearm flexors, rectus femoris and anterior tibialis; among the nerves, vagus, brachial plexus, median, ulnar and sural ones.

Results: We found in all cases increased echogenicity with decreased muscle thickness and fasciculations; these findings allowed us to diagnose the disease in 4 naïve cases not previously identified and later confirmed (Fig. 1 and 2).
Mean diaphragm rest thickness was reduced (2.18 vs 3.45 mm); changes in thickness during inspiration and expiration were also reduced, and lost in the most severe cases (3 pALS) (see Fig. 3).
Median and ulnar nerve cross-sectional area (CSA) resulted smaller in pALS; sural and vagus nerve were spared (Fig. 1 and 2).
Disease severity, measured using Functional Rating Scale (ALSFRS), correlated with quantitative ultrasound data.

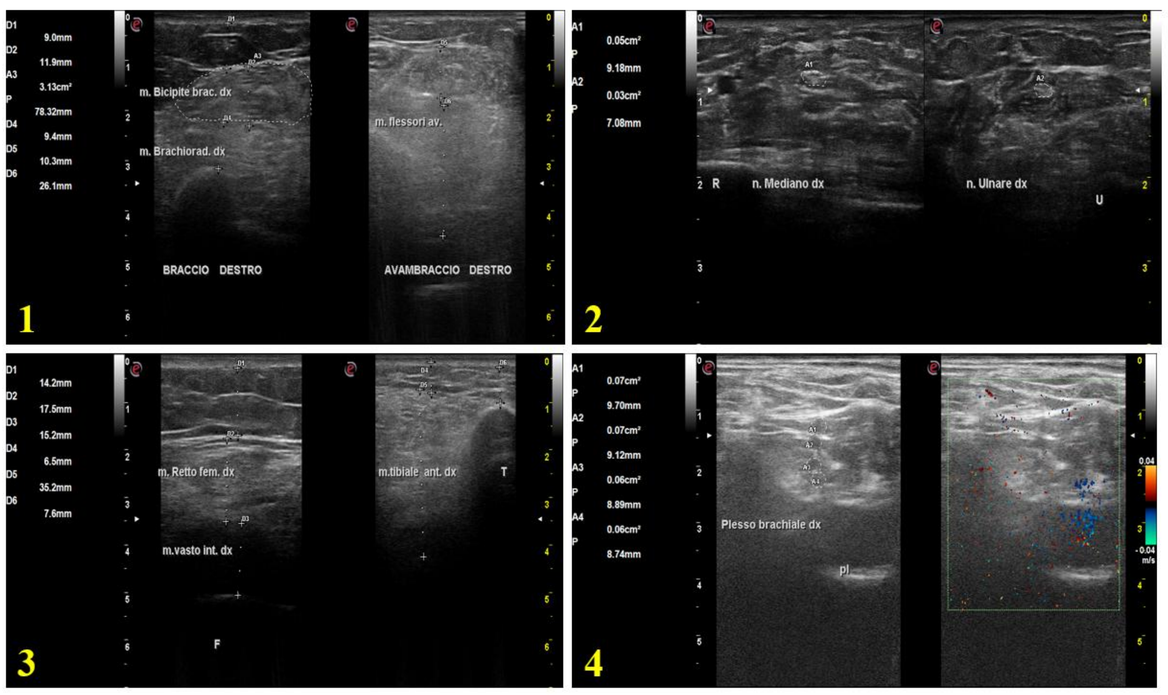

Fig. 1. The muscles (inset 1 and 3), nerves (inset 2) and brachial plexus (inset 4) from a patient with early stage of MND (Case F. P., f, 61 yrs, MND lasting 1 yrs).

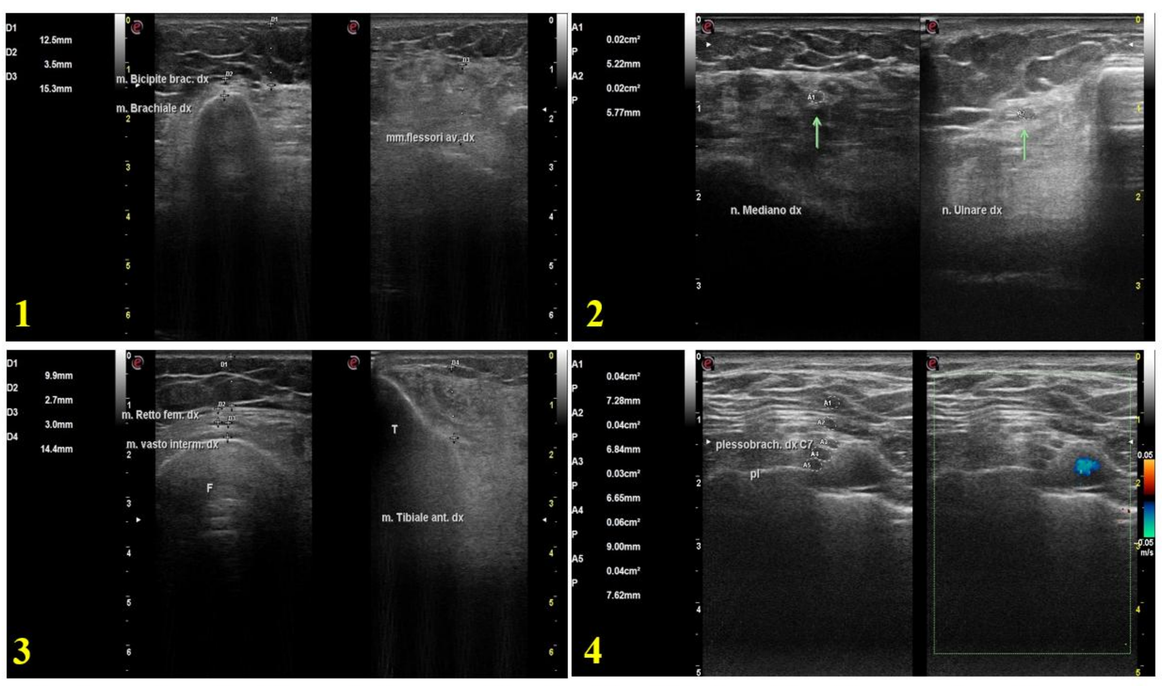

Fig. 2. The abnormal muscles (inset 1; inset 3), nerves (inset 2) and brachial plexus (inset 4) from a patient with severe MND (Case S. P., f, 50 yrs, MND for more than 7 yrs).

Ultrasound of Motor Neuron Disease: *Key Points*

- There are very few studies of nerve ultrasound in patients with amyotrophic lateral sclerosis (pALS), but it does not appear that there is a diagnostically significant change in peripheral nerve caliber or echogenicity in those affected.
- Muscle ultrasound has been studied more often than nerve ultrasound in motor neuron disease.
- In spinal muscular atrophy, ultrasound shows a heterogeneous increase in muscle echogenicity, muscle atrophy, calf hypertrophy, and increased depth of subcutaneous tissue when compared with ultrasound in those with myopathic diseases, and objective gray scale analysis of muscle echogenicity yields a diagnostic sensitivity of 87%.
- Ultrasound can easily detect fasciculations and it allows for painless and rapid scanning over multiple muscle groups.
- Recent studies have shown that with high-resolution ultrasound and proper settings, fibrillations can be detected in those with motor neuron disease.

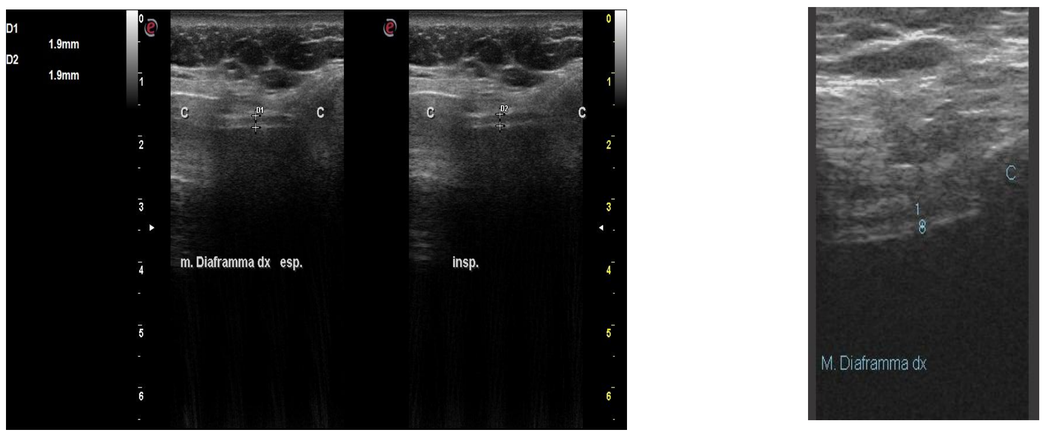

Fig. 3. Abnormal diaphragm seen between the ribs (C), and deep to the abdominal and intercostal muscles during expiration (A) and becoming thicker during maximal inspiration (B). Case S. P., f, 50 yrs, MND from 7 yrs.

On the right side, example of very thin diaphragm Case G.P.P., m, 66 yrs, advanced ALS.

Discussion: Neuromuscular ultrasound resulted a simple, fast and easy, painless and risk-free method, able to provide useful diagnostic as well functional and structural information in pALS. Furthermore, diaphragm US may allow to point out concomitant incoming respiratory failure. Therefore, is desirable that neuromuscular US becomes an essential tool in the diagnostic armamentarium of the neurologist dealing with pALS.

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