Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com

Motor neuron diseases

Motor neuropathies and lower motor neuron syndromes

A. Verschueren

Reference Centre for Neuromuscular Diseases and ALS, University Hospital La Timone, 264 rue Saint Pierre, 13385 Marseille cedex 05, France

INFO ARTICLE

Article history:
Received 16 February 2017
Received in revised form
16 March 2017
Accepted 29 March 2017
Available online 20 April 2017

Keywords:
Motor neuropathy
Lower motor neuron syndrome
Motor neuronopathy
ALS
Spinal muscular atrophy
Benign monomelic amyotrophy

ABSTRACT

Motor or motor-predominant neuropathies may arise from disease processes affecting the motor axon and/or its surrounding myelin. Lower motor neuron syndrome (LMNS) arises from a disease process affecting the spinal motor neuron itself. The term LMNS is more generally used, rather than motor neuronopathy, although both entities are clinically similar. Common features are muscle weakness (distal or proximal) with atrophy and hyporeflexia, but no sensory involvement. They can be acquired or hereditary. Immune-mediated neuropathies (multifocal motor neuropathy, motor-predominant chronic inflammatory demyelinating polyneuropathy) are important to identify, as effective treatments are available. Other acquired neuropathies, such as infectious, paraneoplastic and radiation-induced neuropathies are also well known. Focal LMNS is an amyotrophic lateral sclerosis (ALS)-mimicking syndrome especially affecting young adults. The main hereditary LMNSs in adulthood are Kennedy's disease, late-onset spinal muscular atrophy and distal hereditary motor neuropathies. Motor neuropathies and LMNS are all clinical entities that should be better known, despite being rare diseases. They can sometimes be difficult to differentially diagnose from other diseases, particularly from the more frequent ALS in its pure LMN form. Nevertheless, correct identification of these syndromes is important because their treatment and prognoses are definitely different.

1. Introduction

The motor or motor-predominant neuropathies may arise from disease processes affecting the motor axon and/or its surrounding myelin. Common features are muscle weakness with atrophy that is often distal, and hyporeflexia with no sensory involvement. The disease process also affects the anterior horn cell, namely motor neuronopathy, which is clinically similar but with possible proximal involvement. However, the term 'lower motor neuron syndrome' (LMNS), rather than 'motor neuronopathy', is more generally used. These are rare diseases, but their identification is important when making a differential diagnosis from the more frequent amyotrophic lateral sclerosis (ALS) in its pure LMN form.

Thus, clinical evaluation is an important step for diagnosis. Assessment of disease onset, asymmetrical weakness at onset, distal *vs* proximal weakness, weakness in the distribution of individual peripheral nerves, subacute or progressive

E-mail address: annie.verschueren@ap-hm.fr.
http://dx.doi.org/10.1016/j.neurol.2017.03.018
0035-3787/

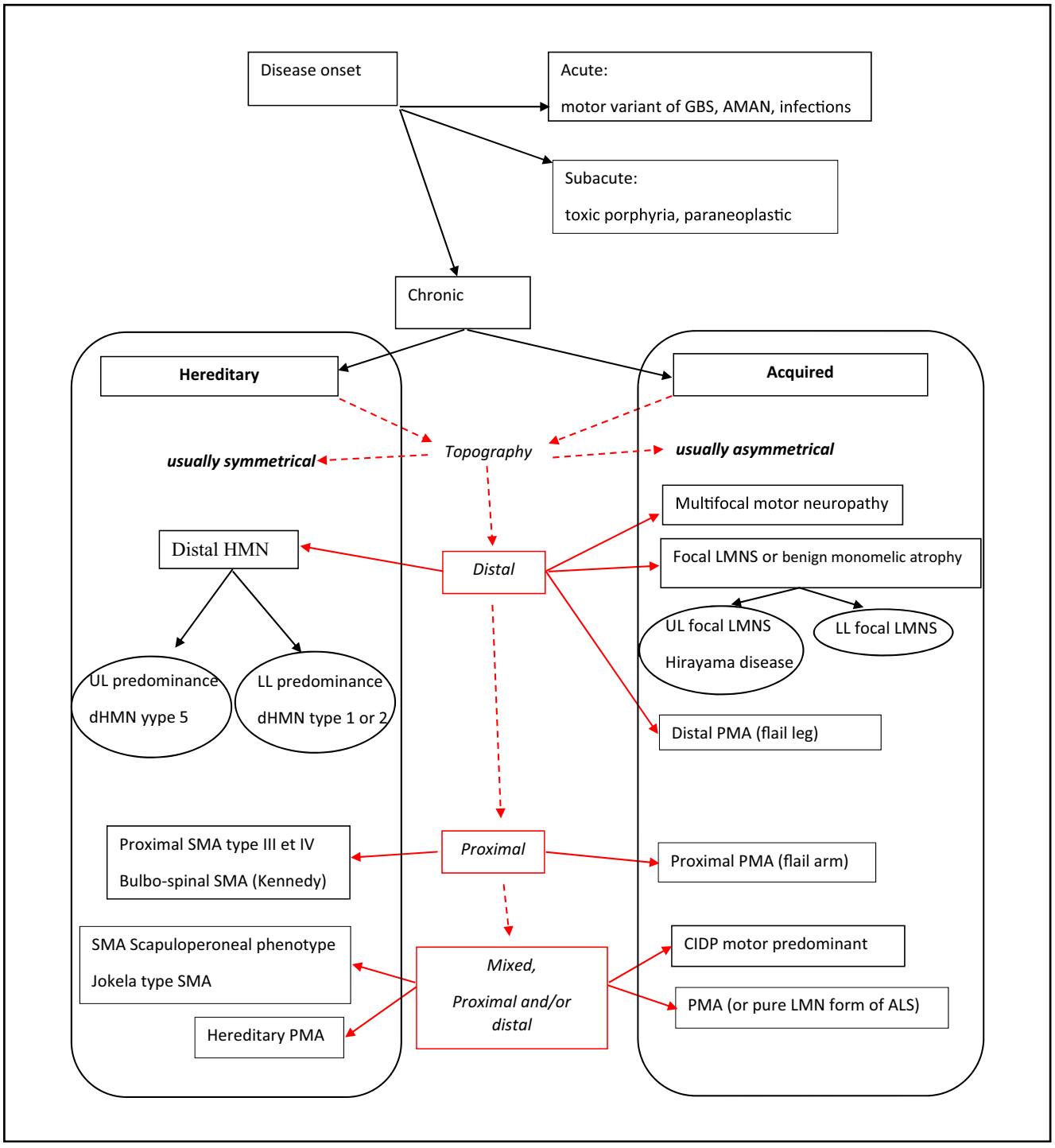

Fig. 1 – Diagnostic algorithm for adult patients with pure motor weakness of peripheral origin, suspected based on signs such as muscular atrophy, hyporeflexia/areflexia, fasciculations and cramps.

onset and stepwise progression should be documented (Fig. 1). The examination should determine the precise pattern of the motor deficit, presence of muscular atrophy or not, twitching, cramps, myokymia and presence or absence of reflexes, all of which are factors that can distinguish the different diseases. Tremor may be present, providing an argument for hereditary disease such as spinal muscular atrophy or spinal–bulbar muscular atrophy.

The patient's medical history (radiotherapy, infections, poliomyelitis and so on) and general examination should be carefully evaluated. The familial history should be done systematically, as it can provide valuable information. For example, distal hereditary motor neuropathy is often autosomal-dominant whereas Kennedy's disease is X-linked. Nerve conduction studies (NCS) and electromyography (EMG) are essential for refining the clinical diagnosis. NCS