Joint Bone Spine 84 (2017) 153–158

Available online at
ScienceDirect
www.sciencedirect.com

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

Review

Neuralgic amyotrophy. An update

Paul Seror

Laboratoire d'électroneuromyographie, 146, avenue Ledru-Rollin, 75011 Paris, France

ARTICLE INFO

Article history:
Accepted 10 March 2016
Available online 2 June 2016

Keywords:
Neuralgic amyotrophy
Plexopathy
Parsonage and Turner syndrome

ABSTRACT

A century after the first description of neuralgic amyotrophy (NA), its pathophysiology remains unknown. An inflammatory (auto)immune pathophysiology is presumed, with mechanical or infectious precipitating conditions, which triggers attacks. Clinically, NA is an acute and painful unique or multiple mononeuropathy that causes palsy, amyotrophy and sensory loss in an asymmetric and patchy distribution. It involves the upper brachial plexus rather than the other parts but also may involve the cervical plexus, lumbosacral plexus and cranial nerves. The impairment can be restricted to one fascicule of one nerve, plexus or root; limited to a few ones; or extensive, involving both upper limbs. Its evolution is usually monophasic and auto-limited and never leads to generalized polyneuropathy. Electrodiagnostically, NA is characterized by severe axonal damage. The recovery is usually good after 6 months to 3 years in 80% of cases. Persistent disability is present in 20% of idiopathic NA cases and is more frequent in hereditary NA, with frequent recurrences, more frequent bilateral impairment, and more atypical distribution (cervical plexus, lumbosacral plexus or cranial nerves) than with idiopathic NA. Hereditary NA is mainly linked to a mutation in the gene of the Septin-9 protein. When the patient is seen early after disease onset, treatment with corticosteroids for 2 weeks seems to shorten the pain duration and the delayed recovery. With diagnosis during the palsy period, treatment is based on pharmacologic and non-pharmacologic therapies according to the complaints of the patient.

1. Introduction

Neuralgic amyotrophy (NA) [1], also called Parsonage–Turner syndrome [1,2] or brachial plexus neuritis [3], is an acute and painful neuropathy that involves mainly the upper brachial plexus. It was first reported after anti-tetanic serotherapy, by Dyke, in 1918. Wyburn-Mason [3] described 42 cases of "brachial plexus neuritis" in 1941, and Spillane [4] reported 46 cases of "localised neuritis of the shoulder girdle" in 1943. Then in 1948, Parsonage and Turner [1] reported 136 cases of "neuralgic amyotrophy". Of note, Dyke, Spillane, Parsonage and Turner were all military medical doctors, which explains the observation of frequent precipitating conditions with the first descriptions.

"Neuralgic amyotrophy", first used by Parsonage and Turner, remains the best term to describe this pathology because it is purely descriptive and presumes none of the pathophysiology nor the exact level of the nerve lesion. It immediately provides the main description of a painful disorder related to a nerve lesion that is severe enough to cause amyotrophy. Other terminologies focus on the region impaired (shoulder girdle) [1,4], the localisation of the nerve lesion (brachial plexus neuritis, plexopathy, neuritis, etc.) [2,3,5,6], or the pathophysiology (immune, idiopathic, hereditary) [7,8]. Since then, some rare series were published that confirmed the previous data or provided some new insights considering diverse clinical, electrodiagnostic (EDX), biological, histological, genetic, imaging and therapeutic data. In 2000, a national expert center for NA was created in Radboud medical university, in The Netherlands.

Since the last reviews considering NA in the French language in 1992 [9] and 2009 [10], the knowledge of NA has progressed. This article revisits this topic with new data from the literature during the last 2 decades.

2. Epidemiology of neuralgic amyotrophy

NA is more frequent in men than women and in the right than left upper limb (both ratios 2 to 1); the disease is asymmetric in 97% of cases. It involves patients from 3 to 80 years old [8]. NA is usually considered a rare disorder (annual incidence 2/100,000 people) [11,12], but recently, an annual incidence of 1/1000 people was reported in primary care, following a specific training of general practitioners on how to diagnose NA (Box 1) [13]. This study confirmed that NA was poorly known and explains why it is diagnosed, in 3 of 4 cases, 28 weeks after disease onset (mean 44 weeks).

E-mail address: paulseror@gmail.com

http://dx.doi.org/10.1016/j.jbspin.2016.03.005
1297-319X/

Box 1: Suggestive symptoms and exclusion criteria for considering a diagnosis of neuralgic amyotrophy [13]

- Neuralgic amyotrophy is probable or definite with:
  - new onset shoulder pain (uni- or bilateral);
  - numeric rating scale score for pain of 7 on a scale of 0–10;
  - abnormal shoulder movement (glenohumeral and/or scapulothoracic) during maximum abduction/anteflexion movement;
  - when first seen 3 weeks after onset: paresis of long thoracic nerve, suprascapular nerve, anterior interosseus nerve.
- Optional signs and symptoms:
  - less severe initial pain with otherwise typical clinical multifocal distribution of weakness and monophasic course;
  - more extensive multifocal paresis of upper extremity(-ies);
  - asymmetric involvement of other upper extremity;
  - areas of hypesthesia and/or paresthesia in the upper extremity;
  - involvement of other peripheral nerves: lumbosacral plexus, phrenic, recurrent laryngeal nerve.
- Neuralgic amyotrophy is excluded with:
  - progression of pain and/or weakness > 3 months (except for pain associated with abnormal compensatory shoulder movements);
  - only passive range of motion constraints in the glenohumeral joint;
  - Horner syndrome;
  - perfectly symmetric weakness distribution;
  - diabetes mellitus.

3. The clinical picture

3.1. Classical clinical pattern

The classical clinical pattern of NA includes 3 successive phases: painful phase, then weakness, amyotrophy and sensory complaints, then recovery [1–4,6,8,9,11,14–18]. These 3 phases are encountered in a similar way with idiopathic NA (INA) and hereditary NA (HNA).

3.1.1. The painful phase

Pain is the first symptom in 90% of cases; it has an acute onset, during the night in 61% of the cases, and is severe, relentless and neuropathic. Usually, pain on a numeric rating scale is greater than 7/10, and involves the shoulder girdle. Its duration varies from 1 day to 2 months; and is longer for males than females (mean 45 vs. 23 days); only 5% of patients have pain for < 24 h and only 10% for > 2 months. After the acute painful phase, 30% to 70% of patients have persistent pain related to diverse causes. Some have neuropathic pain, others develop pain related to muscular compensation required with weakness of palsied muscles, and others have chronic pain without clear significance.

3.1.2. Phase of weakness, amyotrophy and sensory complaints

Weakness may precede the pain in 5% of cases but occurs within 24 h in 34% of cases, after 1–7 days in 39% and after 1–4 weeks in 27%. The amyotrophy appears usually between 2 and 6 weeks and reflects the importance of the axonal loss. It is obvious for superficial muscles (deltoid, supra- and infraspinatus) or is completely invisible for other deep muscles (serratus anterior and pronator quadratus muscles). In descending order of frequency, the most commonly affected muscles are the infraspinatus (72%), serratus anterior (70%), supraspinatus (65%), biceps (61%), rhomboid and pronator teres (53%). The other muscles of the upper limbs are affected in less than 50% of cases, and muscles of the neck (trapezius 20% and sternocleidomastoid 7%) are still less frequently affected [8]. The distribution of the affected muscles corresponds to a root (C5, C6 or C7 more frequently than C8T1), a part of the brachial plexus (the upper and middle plexus more frequently than the lower plexus), a nerve trunk (suprascapular and long thoracic nerves, etc.) or some fascicules of a nerve (anterior interosseous nerve, a motor branch of the median nerve). Lasegue manoeuvre of the upper limb is frequently positive, and frozen shoulder is reported in 17% of cases as a consequence of the NA [8].

Sensory complaints are present in 66% [6] to 78% [8] of cases, were described in the first reports [1,3,4], and appear with the motor weakness and are an integral part of the classical pattern. Their functional consequences are fewer than those of motor weakness and wasting, consequently, NA has been frequently considered by practitioners as a purely a motor pathology. They usually involve the upper cervical root or brachial plexus but may involve some nerve trunks such as the lateral and medial ante-brachial cutaneous nerves, and exceptionally, the distal median or ulnar nerves [16,19,20].

3.1.3. Recovery phase

Overall, 75% of INA cases show good to complete recovery between 6 months and 3 years [2,6,11,14]. In HNA, the multiple attacks of NA (up to 74% of recurrence) increases the risk of incomplete recovery and frequently leads to progressive weakness and disability of one or both upper limbs (20% to 30%) [8]. When the nerve lesion is partial, the recovery is provided by collateral reinnervation and may be completed in 6 to 12 months. In contrast, when the lesion is severe, recovery is based on nerve growth and direct reinnervation, requires 1 to 3 years, and is not always as good. On the whole, the quality and the delay in recovery depend closely on the severity of the axonal loss and the length of nerve growth.

Box 2: Non-exhaustive list of the precipitating conditions in the literature

- Trauma (benign to gunshot wound), exercise, psychological stress, lumbar puncture, cold exposure, burns.
- All kinds of surgery: tonsillectomy, vegetation, appendectomy, hysterectomy, coronary bypass surgery, knee surgery.
- Pregnancy, childbirth and post-partum period.
- Anti-tetanic serotherapy.
- Vaccination: typhoid, diphtheria, tetanus, smallpox, flu, human papillomavirus, etc.
- Bacterial infection: upper respiratory tract infection, typhus, typhoid, malaria, diphtheria, pneumonia, malaria, rheumatic fever, dysentery, sepsis, rickettsia coroni, bartonella henselae (cat claw), borrelia burgdorferi?
- Viral infection: influenza; cytomegalovirus; herpes virus; varicella-zoster virus; parvovirus B19; Epstein-Barr virus; cocksackie virus A2, B; hepatitis B and E virus (10% vs. 0.9% NA among blood donors); Echo 13/30 virus; smallpox; poliomyelitis.

3.2. Precipitating conditions

Precipitating conditions were reported from the very first publication of NA: anti-tetanic serotherapy [15]. Since then, numerous precipitating conditions have been reported (Box 2), in large series and in case reports. Such conditions are common benign trauma, simple or strenuous exercise, pregnancy, any kind of surgery, numerous bacterial or viral infections, and also diverse vaccinations. These conditions are found in 45% to 53% of cases [1,6,8], and occur a few hours to 1 month before the NA attack (within one week 52%) [8,21].