

# A rare presentation of neuralgic amyotrophy in a child and a review of recent literature

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## Abstract

Neuralgic amyotrophy (NA), also known as brachial neuritis and previously known as Parsonage–Turner Syndrome, has an unknown etiology. Patients with NA have a clinical pattern characterized by sudden and acute pain across the shoulder followed by flaccid paralysis. NA has an incidence of one new case per 1000 people per year with an onset of age ranging between 20 and 60 years. We describe a rare presentation of NA in a Caucasian boy who was 11 years old and did not have any other family members affected by NA. All diagnostic studies were normal and he had full recovery 5 months from the onset of symptoms. We revised the recent literature of NA. No specific diagnostic studies can confirm the diagnosis of NA, although magnetic resonance imaging or electrophysiological studies can highlight some special features. Treatment of NA is symptomatic and it is based on analgesic drugs and physical therapy, although early administration of steroids appears to improve the outcome. Prognosis of NA is generally favorable with full recovery usually within 2 years. This disease is typically an adult syndrome, but pediatricians should also be aware of this entity to avoid delays in diagnosis.

## Keywords

Neuralgic amyotrophy, childhood, muscle weakness, pain, magnetic resonance imaging, nerve conduction study, full recovery

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## Introduction

Neuralgic amyotrophy (NA) is a rare syndrome with a characteristic clinical pattern of sudden and acute pain of the shoulder. Pain triggered in specific points worsens with irradiation to the cervical region and to the upper arm. Muscle weakness and flaccid paralysis of the shoulder girdle and upper arm, frequently without sensitivity deficit, appear later.<sup>1,2</sup> NA is a disabling disease that usually affects people aged between 20 and 60 years with slight male predominance (male: female = 2.3:1)<sup>3</sup> and its clinical presentation may mimic other pathologies.<sup>4</sup> NA is the most common cause of nontraumatic brachial plexopathy with an incidence of one case per 1000 people per year.<sup>5</sup> However, in recent years, the concept of plexopathy is changing to multiple peripheral mononeuropathy.<sup>6</sup>

The exact cause of NA is still unknown. Some authors hypothesize a viral etiology, while others have suggested a role of various infections as triggers of onset. Other possible hypotheses include a recent history of vaccinations, local surgery, local traumas, or systemic diseases with concomitant nerve damage involved.<sup>7</sup> The main pathophysiological hypotheses are an interaction between organ-specific autoimmune triggers and mechanical vulnerability in patients with genetic predisposition.<sup>8</sup> No diagnostic exams provide a specific result for diagnosis, although sometimes magnetic resonance imaging (MRI) of the shoulder can highlight signs of neuritis.<sup>6</sup> Therefore, the diagnosis of NA is achieved by a careful history and examination excluding other pathologies. There is often no positive history of inflammatory or infectious diseases with NA. Imaging studies, such as shoulder and cervical spine radiographs, and brachial plexus and cervical MRI are helpful in excluding any associated local pathological processes.<sup>5,9</sup> Routine nerve conduction studies (NCSs) usually cannot prove NA

because it may appear to affect muscles of the C5 root, the upper trunk of the brachial plexus, and axillary and musculocutaneous nerves. Conduction studies of axillary nerve compound muscle action potentials (CMAPs), musculocutaneous nerve CMAPs, and lateral antebrachial cutaneous nerve action potentials can often detect an abnormal amplitude reduction in patients with NA.<sup>10</sup> These studies can also help to avoid any unnecessary surgical intervention and identify any coexisting neurogenic conditions, if any doubt persists.<sup>11</sup>

No specific treatment has yet been proved efficacious in NA,<sup>12</sup> but non-steroidal anti-inflammatory drugs can be useful to relieve pain, mainly in the early stages. Use of corticosteroids is not yet fully accepted.<sup>12</sup> Some authors recommend immobilization of the affected upper arm to relieve the pain and to prevent stretching of the affected muscles.<sup>8</sup> Physical therapy for NA is also recommended.

The prognosis of NA is currently recognized as not excellent; it is widely variable from an early recovery within 1 month to no observable recovery after several years.<sup>13</sup> In this report, we describe an unusual presentation of NA in a child who had a full spontaneous recovery.

## Case report

A healthy Caucasian boy who was 11 years old presented with a spontaneous sudden pain that was localized in his right shoulder and unilateral upper arm (right hand was spared) in the absence of trauma. His family doctor prescribed paracetamol, but there was no relief of the pain. Therefore, he visited the emergency department where shoulder and upper arm radiographs show no fractures. However, an orthopedist recommended dressing and immobilization of the arm for 2 weeks. Ten days later, because of constant and persistent pain, the family

doctor requested some laboratory tests. A blood cell count, levels of C-reactive protein, creatine-phosphokinase, and lactate dehydrogenase, urinalysis, and culture of a pharyngeal sample were normal. When the orthopedist removed the dressing, the boy presented with considerable weakness in the upper arm. The family doctor then recommended a pediatric rheumatological examination. There was pain to acupressure in the low cervical area with insertion and along the ridge of the right biceps with strength deficit of this muscle. This specialist doctor prescribed ex-juvantibus therapy with gabapentin 300 mg three times a day for 3 weeks as a neuroprotective action. The severe pain persisted and the muscle weakness worsened until appearance of paralysis. At this time, pain and muscle weakness also appeared on the right hand and his family doctor recommended hospitalization.

In our Pediatric Unit, the boy presented with an antalgic posture of the right arm that appeared adduct and outstretched with severe pain at acupressure of the right trapezium muscle, shoulder, and elbow. We gradually withdrew gabapentin because of problems of concentration as reported by the mother and we prescribed ibuprofen 10 mg/kg three times a day.

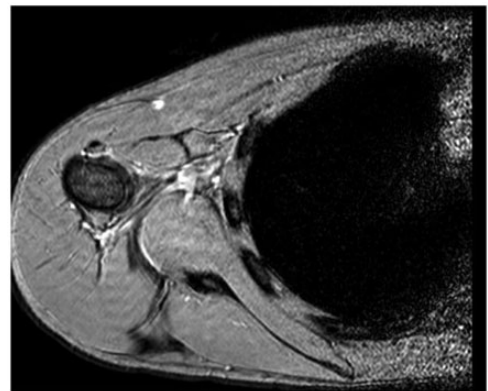
A neurological examination showed normal sensory function, no cranial nerve deficit, no neural sensitivity defects, no motor deficit in the lower limbs, normal tendon reflexes, and absence of Babinski sign. Muscle weakness was apparent only in the right arm with a functional limitation. In particular, strength deficit of the biceps and flexor digitorum muscles was observed, suggesting a median nerve deficiency. No signs of deficiency in other muscle groups were evident, although the intense acupressure pain made accurate assessment difficult. Cerebellum-vestibular tests were negative, except for the index finger-nose test on the right side because of pain. The neurologist (an adult

neurologist) mentioned an adult syndrome with the same symptoms as those seen in our child called NA, but she had never seen NA in children.

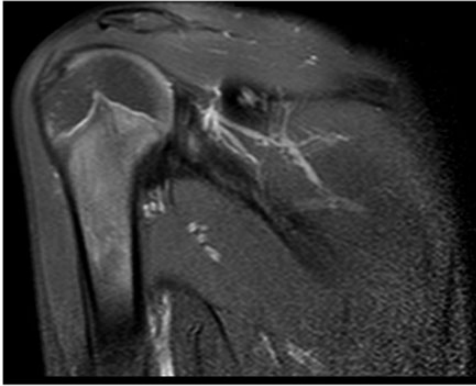
We then repeated blood examinations (complete blood count, C-reactive protein, creatine-phosphokinase, lactate dehydrogenase, serum protein electrophoresis, rheumatoid factor, and anti-nuclear, anti-nucleolar, anti-neutrophil cytoplasm and anti-Borrelia antibodies), which were normal. The orthopedist repeated shoulder and cervical radiographs, which were negative.

The shoulder and cervical MRI, performed to rule out expansive lesions of the cervical spine and cervical hernias, showed no abnormalities. In particular, MRI showed no signs of neuritis with regular signal intensity of bone parts of the shoulder, regular rotator cuff tendons, and normal tropism of the shoulder girdle muscles (see Figures 1, 2, and 3). There was normal signal intensity of examined vertebral metamer (from C1 to T8) and of the spinal cord.

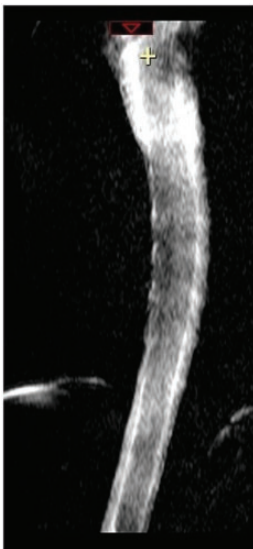
Then, NCSs and electromyographic studies have performed. No brachial plexus lesions were observed. Therapy with steroids and vitamin B6 was recommended by the



**Figure 1.** A transverse magnetic resonance imaging scan of the right shoulder shows no sign of neuritis.



**Figure 2.** A sagittal magnetic resonance imaging scan of the right shoulder shows no sign of neuritis.



**Figure 3.** A magnetic resonance imaging scan of the cervical spinal cord shows no sign of neuritis.

neurologist. In particular, NCSs showed normally evoked compound muscle action potentials and sensory nerve action potentials with preserved conduction velocity, distal latency, and amplitudes. There was symmetrical evocability of the F wave as shown by analyzing the sensory ulnar and median bilateral nerves and circumflex

nerve, the deltoid muscle, the right motor ulnar and median nerves, stimuli at the wrist, elbow, arm, and axilla, the left ulnar and median nerves from elbow to wrist, in the ulnar and musculocutaneous nerves, bilateral biceps muscle, and Erb's point. Needle electromyography showed no pathological resting potentials by analyzing the supraspinatus, deltoid, triceps, biceps, extensor index, and thenar and hypothenar eminence. An increased insertion activity of the needle was observed and voluntary activity of the muscles was poorly assessable because of antalgic contraction. Furthermore, motor unit action potentials were of normal amplitude, morphology, and duration. At this time, therapy with steroids and vitamin B6 was recommended by the neurologist.

A physiatrist assessed all exams and symptoms and suggested the diagnosis of brachial plexopathy or NA. None of the patient's family members ever had similar symptoms. Approximately 4 months after the onset of symptoms and after intensive bilateral physical therapy, the boy no longer had any symptoms. A slight strength deficit was still present in the right biceps and in the right flexor digitorum muscles. Five months after onset, he had complete regression of clinical signs.

Written informed consent for publication was obtained from a parent of the patient.

## Discussion

The exact cause of NA is unknown and it is often unrecognized by doctors, with an average delay of 3 to 9 months before its diagnosis.<sup>7</sup> Many factors have been proposed in the etiology of NA, such as trauma, heavy exercises, infections, viral diseases (Coxsackie B virus, hepatitis E virus), recent vaccinations, recent local surgery, and autoimmune diseases.<sup>5</sup> Hepatitis E virus was recently recognized to contribute to NA.<sup>14</sup> Although NA is rare in children, it should be considered in differential

diagnosis. A rare hereditary form of NA has been previously described.<sup>8</sup> In our case, there were no affected family members and the mother refused to have genetic research performed, preferring to postpone it to a possible future attack.

Characteristically, NA begins with acute pain in the shoulder, followed by profound muscle weakness at the upper unilateral arm.<sup>12</sup> This pain is acute, often severe, and throbbing, irradiating from the shoulder distally to the arm or proximally to the neck. This particular clinical course of symptoms (severe pain followed by muscle weakness) helps with diagnosis of NA. NCSs are useful in ruling out a traumatic injury, nerve compression, or neural trunk lesions.<sup>13</sup> NA conduction studies of the involved nerves can often detect an abnormal reduction in amplitude, but in our case, NCSs were normal. Therefore, in our opinion, NA remains the only plausible diagnosis based on the patient's symptoms, his clinical features and his clinical history and course with full recovery. We did not determine if hepatitis E virus was present because it is unusual in Italy.

In a recent review, Al-Ghamdi and Ghosh<sup>1</sup> analyzed 22 patients aged between 6 and 18 years old who were affected by NA. Pain was the clinical sign observed in all of the patients with muscle weakness of the periscapular and scapular winging in 6 and 13 patients, respectively. In two children, a viral infection preceded the onset of symptoms. Electromyography, which was performed in 21 patients, showed sensory nerve abnormalities in 5 children, and the serratus anterior muscle was the most commonly affected area. The treatment was mainly supportive, even if 4 patients received immunotherapy. Persistent pain and residual motor deficits were observed in most children during a 36-month follow-up.<sup>1</sup> Høst and Skov<sup>3</sup> analyzed 58 pediatric cases of idiopathic NA. Pain was present in 47% of patients, whereas it was

not present in 25% (unknown in other patients). The right and left sides appeared involved in 74% of patients, and in 26%, localization of the pain was unknown. In two cases, bilateral plexus involvement was observed. With regard to prognosis, 63%, 25%, and 13% of patients had full, partial and no recovery, respectively. Full recovery was obtained after 7.9 months, partial recovery was obtained after 17.4 months, and for patients who obtained no recovery, the follow-up time was 14.6 months.<sup>3</sup> Mrowczynski et al.<sup>15</sup> summarized cases of infant NA and they concluded that the most common presenting symptoms were single arm immobility and pain (such as our patient), while the most common treatment was to watch and wait. Most patients' symptoms gradually resolved over a period of months to 1 year.

Because of the lengthy, but common delay in diagnosis of NA, systematic treatment with prednisolone is rarely recommended. However, van Eijk et al.<sup>16</sup> empirically administered prednisolone treatment in the early stages of NA in adults while they were still suffering from severe pain either with or without conventional analgesic measures in an open-label study. They showed that corticosteroid treatment in the early stages of a NA attack can positively affect outcome. With regard to the untreated patients, a significantly higher proportion of patients who received oral prednisolone recovered early from their paresis. When taken in the first month, prednisolone decreased the average duration of the initial pain (20.5 days in the historical controls and 12.5 days in the study group). Therefore, these authors recommended prednisolone, in the absence of contraindications, for early treatment of patients with acute NA. Although side effects occurred in 20% of their patients, they never had a reason to discontinue treatment. The effect of early administration of prednisone was confirmed by a



recent Cochrane review.<sup>7</sup> When patients were analyzed from 1966 to 2009, this review showed that no randomized, controlled trials were performed to evaluate treatment of NA. The authors of this review concluded that using oral prednisone, in the first month of symptom onset, may increase the speed of recovery.

In conclusion, NA is often a self-limited disease and the main treatment remains a conservative approach. Analgesic drugs (non-steroidal anti-inflammatory drugs) are required during the early stages of the disease because of severe pain,<sup>1</sup> but these drugs and physical therapy (massage, ultrasound of electrical stimulation therapy, and rehabilitation) do not appear to improve the time to functional recovery. Oral prednisone provided in the first month after onset appears to decrease the duration of symptoms.<sup>7</sup> We consider that we obtained a full recovery of functionality of the upper arm in our patient possibly because of his young age. Children have enormous potential to recover from possible damage suffered. Finally, although typical of adulthood, even pediatricians should be aware of NA to avoid diagnostic delays. The patient's history and a thorough clinical examination remain the cornerstones of diagnosis of NA.

### List of abbreviations

NA: neuralgic amyotrophy; MRI: magnetic resonance imaging; NCSs: nerve conduction studies

### Availability of data and material

Data sharing is not applicable to this article because no datasets were generated or analyzed during the current study.

### Authors' contributions

MM conceived the case report and wrote the manuscript together with SI. VM collected the recent studies in literature. PG and GG critically

revised the manuscript. MM and SS corrected the revised versions.

### Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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