Um Caso Atípico e Grave de Amiotrofia Neurálgica
An Atypical and Severe Case of Neuralgic Amyotrophy

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Resumo
A amiotrofia neurálgica ou síndrome de Parsonage-Turner é uma síndrome incomum, de etiologia desconhecida, tipicamente caracterizada pelo início súbito de dor nos ombros e/ou membros superiores seguido de défices neurológicos progressivos. Um processo imune/autoimune parece estar na origem desta síndrome. A amiotrofia neurálgica é um diagnóstico clínico, no entanto, os exames complementares podem ajudar a confirmar a suspeita clínica e a excluir outras patologias. O tratamento consiste predominantemente no controlo da dor e reabilitação. Os pacientes com amiotrofia neurálgica podem ser avaliados por um vasto número de clínicos na fase inicial da doença antes de ser efetuado o diagnóstico correto. Este atraso é resultado dos diferentes fenótipos que esta síndrome pode exibir, complicando o diagnóstico. É importante que os clínicos tenham presentes as manifestações clínicas clássicas que geralmente seguem um curso característico com início abrupto de dor na cintura escapular seguido de déficits neurológicos progressivos como fraqueza muscular, amiotrofia e alterações sensitivas.

Palavras-chave: Dor de Ombro; Neurite do Plexo Braquial.

Abstract
Neuralgic amyotrophy or Parsonage-Turner syndrome is an uncommon disorder, of unknown etiology, typically characterized by abrupt onset of shoulder and upper extremity pain followed by progressive neurologic deficits. An immune/autoimmune process seems to have more support in development of neuralgic amyotrophy. Neuralgic amyotrophy is a clinical diagnosis, however, further diagnostic studies can confirm clinical suspicion and help exclude other causes. The treatment consists predominantly in pain control and rehabilitation. Patients with neuralgic amyotrophy, can be evaluated by a wide range of clinical specialists in the early stage of disease before a correct diagnosis be made. This is a result of different phenotypes that this disorder exhibits, that complicate the diagnosis. It is important for clinicians to beware of the classic clinical manifestations that usually follow a characteristic clinical course of abrupt onset of shoulder girdle pain followed by progressive neurologic deficits as muscle weakness, amyotrophy and sensory abnormalities.

We report the case of a 32-year-old man who had a severe bilateral and atypical shoulder involvement, that made the most probably diagnostic not so evident.

Keywords: Brachial Plexus Neuritis; Shoulder Pain.

Introduction
Neuralgic amyotrophy (NA) is an uncommon disorder, with high clinical variability, typically characterized by abrupt onset of shoulder and upper extremity pain followed by progressive neurologic deficits.

Although this clinical condition is most commonly referred to as Parsonage-Turner syndrome (PTS), NA is actually the most common term in the literature and probably the best term to describe this syndrome because as some authors argue “it’s purely descriptive and presumes none of the pathophysiology nor the exact level of the nerve lesion”.

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The incidence of idiopathic form is estimated at two to four per 100 000 per year, but recent studies show that this can be significantly higher.

Electrodiagnostic studies are extremely useful in localizing the lesion and confirming the diagnosis.

There is no proven specific treatment that decreases neurological damage or improves prognosis. It is a self-limited illness, and patients could have full restoration of function but recovery is usually prolonged, months or even years, and can be incomplete.

We present a case of a patient who had NA with severe bilateral shoulder involvement.

**Case report**

A 32-year-old right-handed man was referred to our neurology department five days after the onset of severe bilateral shoulder pain and inability for shoulder abduction. He had no history of recent surgery, infection, immunization or trauma, but he reported having helped carry a coffin at a funeral 48 hours before the beginning of pain. The pain, described as “burning”, woke him up during the night, began in the left shoulder and after one hour appeared also in the right shoulder, with maximal intensity a few hours later (NPRS - Numeric Pain Rating Scale of 8/10). Within the next eight hours, he noted a progressive inability to perform shoulder abduction. He went to an emergency department and then to his family physician, before the last one referred him to our hospital. In both times, he was medicated with painkillers with slight improvement of complaints. His past medical history included diabetes mellitus (DM) diagnosed 10 years before, treated with antidiabetic medication (metformin 1000 mg + vildagliptin 50 mg twice a day and glicazide 60 mg once a day) and On his initial presentation, he reported bilateral shoulder pain radiating from the neck, an inability to shoulder abduction and paresthesia in the lateral aspect of right shoulder and left thumb. He denied constitutional symptoms like fever or malaise or other systemic signs/symptoms and did not describe Lhermitte’s sign.

Physical examination revealed no changes on inspection, marked weakness on shoulders abduction/anterior elevation - grade 1 on shoulder abduction/anterior elevation/external rotation bilaterally on Medical Research Council (MRC) scale - and slight bilateral elbow extension weakness, grade 4 on MRC scale bilaterally, with complete passive range of motion (ROM) on shoulder and upper limbs joints (Fig.s 1 and 2). He had hypoesthesia on the lateral aspect of the right shoulder and deep bicipital tendon reflexes were absent. No more significant changes were on musculoskeletal/neurologic examination were found (including clinical signs of polyneuropathy).

Routine laboratory tests were normal, except hyperglycemia (274 mg/dL). Cervical computed tomography (CT) scan did not show significant abnormalities. So, he was admitted to our Neurology department for further investigation and therapeutic orientation.

During the first days we had some difficulty in pain control. Pain was partially improved using tramadol 50 mg three times a day and 3 mg morphine as needed. We proceeded with etiological investigation. The cervical magnetic resonance imaging (MRI) had no significant abnormalities too.

He took new blood samples for serologic and autoimmunity tests, inflammatory, thyroid and viral markers, folic acid and B12 vitamin. All results were normal, except glycated hemoglobin (HbA1c) that was 9.4%.
He was evaluated by an endocrinologist due to his early onset of DM and azoospermia of unknown etiology. He began insulin therapy and a later reevaluation was scheduled.

Thirteen days after the onset of symptoms we performed an electromyography/nerve conduction study (EMG/NCS) that showed evidence of acute denervation in muscles supplied by the suprascapular and axillary nerves bilaterally.

After this, the diagnosis of NA seemed more likely. We decided to complete the study with a cerebrospinal fluid analysis, that showed normal results except for elevated proteins (1.02 g/L) and glucose (132 mg/dL).

We decided to start treatment with methylprednisolone 1gr intravenous during five days and also gabapentin, progressively, until 300 mg three times a day. In the day after the first dose of corticosteroid his pain improved significantly (NPRS from 7/10 to 2/10). During the hospitalization period the bilateral proximal brachial paralysis stayed mostly unchanged, with only slight improvement at discharge (MRC grade 2 on shoulder abduction/anterior flexion bilaterally). He was evaluated by Physical and Rehabilitation Medicine (PRM) three days before hospital discharge and referred to a rehabilitation program.

Before discharge from the hospital we repeated the EMG/NCS (22 days after the onset of symptoms) which showed signs of acute denervation in the deltoids and infraspinatus muscles bilaterally, with normal biceps and rhomboids major muscles, suggestive of recent severe axonal injury with a probable location at level of the upper primary trunks of brachial plexus/C6 spinal root bilaterally.

The acute onset of pain followed by neurologic deficits and the electrodiagnostic results, were deemed compatible with the diagnosis of NA, in a man with nervous susceptibility to noxious stimuli due to DM. The effort of carrying a coffin for a long period of time, a few days before, was possibly the triggering factor.

At discharge, he continued medicated with insulin 32 + 18 units, same dose of gabapentin and tramadol 50 mg twice a day. One week later, he began physical therapy and kept the pharmacologic treatment. He was advised not to lift up considerable weights to prevent shoulder subluxation.

As he worked as an office employee, he returned to work when the pain subsided. He returned four weeks later to PRM consultation (eight weeks after the onset of symptoms), at the time presenting no pain complaints, having even suspended all painkiller medication by his own will since the previous two weeks, but bilateral proximal brachial paralysis was still present, without changes in muscle strength since the last evaluation (Figs. 3, 4 and 5).

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The rehabilitation program lasted for eight months. During the first phase of rehabilitation program, we focus in pain control, maintenance of full ROM and...
avoid amyotrophy of the affected muscles. For that, we use electrotherapy, manual therapy techniques as passive glenohumeral mobilizations and myofascial release. It was used electric muscular stimulation to slow the progression of atrophy, reduce fibrosis and improve the circulation and nutrition of the affected muscles. TENS (transcutaneous electrical nerve stimulation) was also used in pain management. Scapular stabilization exercises were also started at this stage. The timing and the role of strengthening exercises were primarily dependent on the degree of muscle denervation and weakness.

When the pain disappeared, the treatment continued to focus on ROM exercises and scapular stabilization, and progressively began strengthening of the rotator cuff and deltoid muscle, as strengthening of the muscles of the upper limb.

Patient’s education about energy conservation strategies and importance of good ergonomics had an important role in the rehabilitation process.

A year later, he maintains regular consultations by PRM. In last evaluation he had full active ROM of shoulders and reported no pain. He maintained a slight decrease in muscle strength in right shoulder abduction, anterior elevation and external rotation.

Discussion

NA is an acute and painful neuropathy that involves mainly the upper brachial plexus, which the exact pathophysiology remains unknown; but a combination of underlying genetic predisposition, susceptibility to mechanical injury of the brachial plexus and an immune/autoimmune trigger seem to have the most support at the moment.1 NA has been linked to prior events or diseases, such as trauma, viral infections, immunizations or heavy exercise.4 This last one, we suspect that was the “trigger” in our patient.

Due to high variability of clinical presentation and lack of knowledge by the clinicians, NA diagnosis is often delayed, being diagnosed as shoulder or cervical disorders.3 The possible association between DM and other disorders with NA is uncertain.4

There is a genetic form of NA that is characterized by recurrent attacks of pain. It occurs ten times less frequently than its idiopathic counterpart.1

Commonly, the characteristic clinical presentation is an acute, severe neuropathic pain in the shoulder and upper extremity lasting for several days or weeks, followed by muscle weakness, amyotrophy and sensory loss as the pain diminishes/disappears.8 The beginning of pain, with neuropathic characteristics that awakes the patient during the night and reaches its maximum a few hours later is quite common. Pain is the presenting and predominant symptom in more than 90% of patients, but usually subsides in a few weeks.8

In the early stage it may be difficult to distinguish whether the inability to actively mobilize the shoulder is due to intense pain or weakness. The short time between the onset of pain and muscular weakness, as well as the severity of its attainment, makes early diagnosis difficult, as, in up to one third of all cases, paresis begins in the first 24 hours after pain.8

NA most commonly involves suprascapular, long thoracic and axillary nerves individually or in combination,1,3 so, it will affect predominantly shoulder abduction, anterior elevation and external rotation. Bilateral involvement, usually in an asymmetrically way, may be present in up to 30% of cases.4 In our patient, suprascapular and axillary nerve were involved bilaterally, supported by clinical examination and EMG/NCS, and, it clinically seemed that left long thoracic nerve was also affected, however this was not evaluated in electrodiagnostic studies (Fig. 4).

No currently available tests can unequivocally confirm or exclude NA itself. Blood examinations and CT or MRI serve mainly to exclude other disorders.1 MRI of brachial plexus can show signal change consistent with inflammation of the brachial plexus,11 or muscle changes, but, most of the time, MRI is normal.8 Cerebrospinal fluid abnormalities, consisting of increased protein are occasionally reported in NA, as in our patient.1 Electrodiagnostic studies are of great value for the diagnosis. Usually, presents as an axonopathy, and, due to the different forms of involvement, it may be necessary to evaluate several muscles that are not routinely tested.13

The treatment consists predominantly in pain control and rehabilitation, ideally with a multidisciplinary approach, including intervention of physiotherapy and occupational therapy, coordinated by a physiatrist. In the first phase, the hyperalgic phase, analgesia, relative rest of the limb and maintain shoulders ROM are recommended. The use of corticosteroids is controversial.1 In our patient, a five day course of 1gr intravenous methylprednisolone was performed, with a significant pain reduction after the first dose. We also prescribed him gabapentin, but there is no consensus about its benefit in the early stages due to its long latency period until the beginning of its effect.

After the pain has improved, the second phase of rehabilitation can be started which aims to continue reestablish/ maintain the ROM, followed by muscle strengthening. A good and progressive rehabilitation program is crucial to avoid complications. During the rehabilitation process, one of the main goals in our patient was to avoid glenohumeral subluxation since...
almost every rotator cuff muscles and deltoid were affected. Patient was advised to avoid lifting heavy objects and to use an orthosis for centering humeral head on the right shoulder. Once DM and immobilization are two known risk factors for the development of adhesive capsulitis, we began early shoulder mobilizations trying to avoid it.

Recovery differs between patients, with more than a half of them are left with residual paresis and exercise intolerance, especially of the periscapular muscles. This can lead to biomechanical changes which will increase the risk of joint pathology and strain of the paretic and compensating muscles.1, 4, 8, 10

This case shows the importance of a detailed clinical history since NA diagnosis is predominantly clinical. As result of different phenotypes that this disorder could present, it is important for clinicians to know the classic clinical manifestations of this syndrome, but be aware that there are different forms of presentation and severity.

Referências / References: