Respiratory Medicine Case Reports 28 (2019) 100913

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

A strange case of dyspnoea

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ARTICLE INFO

Keywords:
Dyspnoea
Parsonage-turner syndrome
Neuralgic amyotrophy

ABSTRACT

Acquired neuralgic amyotrophy, described for the first time by Parsonage and Turner, is a rare idiopathic disease that may occur in otherwise normal healthy individuals. It typically begins with sudden, unilateral shoulder pain that may also involve the neck and/or arm. Less frequently, the disease involves nerves other than those of the brachial plexus, such as phrenic nerves, resulting in dyspnoea. The diagnosis is based on the clinical presentation and is generally supported by electroneurography/electromyography. We report the case of a 45-year-old white man who was referred to our clinic for acute dyspnoea preceded by severe neck and right shoulder pain. Corticosteroid therapy ameliorated the clinical picture, but without a complete recovery.

1. Introduction

In 1948, Parsonage and Turner described a neurological syndrome that became increasingly common during WWII, characterised by sudden shoulder pain followed by atrophic paralysis in the muscles of the shoulder girdle, in some cases associated with sensitivity alterations. The authors suggested the name “neuralgic amyotrophy” (NA) [1]. Here we described a case of NA onset as acute dyspnoea.

2. Case report

We report the case of a 45-year-old white man who was referred to our clinic for acute dyspnoea preceded by severe neck and right shoulder pain with no history of trauma.

The patient had hypertension treated with losartan 100 mg/day and his family history was unremarkable for respiratory, neurologic and autoimmune diseases. Blood gas analysis showed a slight pO_2 reduction, with normal pH and pCO_2. Complete blood count, CRP and D-dimer were in the normal range. A chest X-ray (Fig. 1A) showed right hemidiaphragm elevation. These findings could not fully explain the clinical picture, so a CT scan of the thorax, abdomen and pelvis was performed, which confirmed the right hemidiaphragm elevation without other significant alterations. In order to clarify the origin of this picture, a cervical spine MRI scan was performed that showed multiple spinal disc herniations extending from C4 to C7 with no spinal cord alterations. Finally, electroneurography/electromyography (ENG/EMG) was performed. ENG showed an increased duration of the right compound muscle action potential of the phrenic nerve, suggesting a demyelinating nerve injury (Fig. 1B), while needle EMG of the right hemidiaphragm revealed signs of denervation. Together, these data were indicative of a partial lesion of the right phrenic nerve and led us to make a diagnosis of acquired NA. The patient received corticosteroid therapy with prednisone 0.5 mg/kg/day for 15 days and respiratory rehabilitation therapy with progressive clinical improvement, but without a complete recovery.

3. Discussion

Acquired NA is a rare idiopathic disease that more frequently affects male adults in the second or third decade of life with an incidence estimated at 2–4/10^5/year. There is also an inherited form of the disease caused by mutations in the SEPT9 gene [2].

NA may occur in otherwise normal healthy individuals and typically begins with sudden, unilateral shoulder pain that may also involve the neck, scapula, arm, forearm or hand and that often wakens the patient in the middle of the night or early morning. The pain lasts for a few days or weeks, rarely with a longer duration, and while it subsides a patchy paresis of the muscles innervated by the brachial plexus nerves arises, associated with muscle atrophy and in some cases with sensitivity alterations such as hypoesthesia. Less frequently, the disease involves nerves other than those of the brachial plexus, such as phrenic nerves, resulting in dyspnoea [2,3].

Recent viral infection is the most common precipitating condition that has been associated with NA, so it has been theorised that a viral

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https://doi.org/10.1016/j.rmcr.2019.100913
Received 15 May 2019; Received in revised form 21 July 2019; Accepted 21 July 2019
Available online 22 July 2019

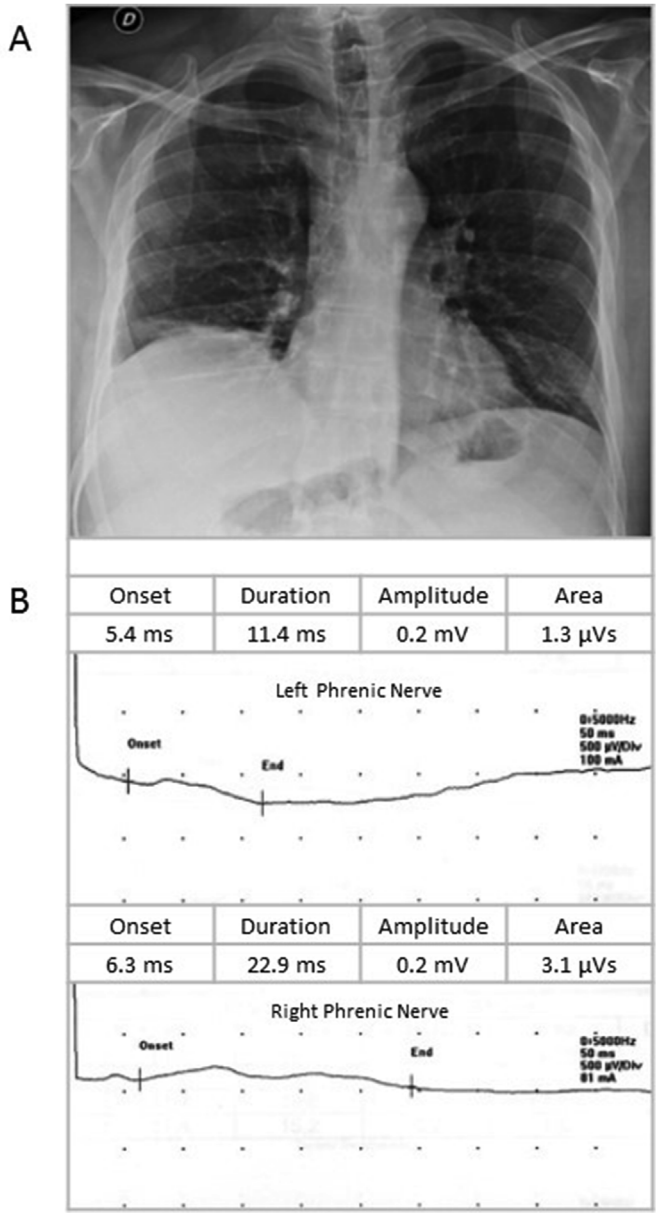

Fig. 1. A) Chest X-ray showing right hemidiaphragm elevation. B) Electroneurography of both phrenic nerves showing an increased duration of the right compound muscle action potential.

illness may directly involve the brachial plexus or that it could ignite an autoimmune response. Other factors that may be associated with NA are drug use, heavy exercise, trauma, surgery and rheumatic diseases. The reason why certain nerves are affected more frequently is not completely understood.

The diagnosis is based on the clinical presentation, in particular when there is the involvement of different nerves that do not follow a common root or plexus pattern. Other diseases similar to NA should be considered and excluded, such as radiculopathy. In the latter, pain varies with posture, worsens with the Spurling test, improves with immobilisation and is localised in the same dermatomes of sensory and motor symptoms. MRI and ENG/EMG may be useful, but generally the latter is the only imaging modality that positively supports the diagnosis. Chest X-ray may be useful to rule out a Pancoast tumour and sometimes to show diaphragm elevation if the phrenic nerve is involved [2–4].

Treatment of the acute phase consists of pain management with NSAIDs, opiates and neuroleptics; physical therapy also plays an important role. Steroids may shorten the duration of the disease, but there is no strong evidence supporting such treatment [3]. Patients with NA usually recover spontaneously, but symptoms may last for many months or years and in some cases complete recovery will never occur [5].

Conflicts of interest

The authors declare that they have no conflict of interest.

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