International Journal of Healthcare Sciences ISSN 2348-5728 (Online)
Vol. 5, Issue 1, pp: (116-121), Month: April - September 2017, Available at: www.researchpublish.com

An Updated Review in the Management of Diaphragm Paralysis Related to Red Cross Syndrome: Systematic Review

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Abstract: **The diaphragm, the most essential muscle of ventilation, develops negative intrathoracic pressure to initiate ventilation. Innervated by cervical motor nerve cells C3-C5 via the phrenic nerves, these two nerves offer both sensory and motor function to the diaphragm. With contraction, the cone-shaped muscle of the diaphragm decreases intrapleural pressure during inspiration and consequently helps with motion of air into the lungs. Diaphragmatic paralysis is an unusual, yet underdiagnosed reason for dyspnea.We provide 3 cases of symptomatic diaphragm paralysis where a clear clinico-pathologic diagnosis could be determined, specifically a vascular compression of the phrenic nerve in the neck caused by an adherent or tortuous transverse cervical artery. We suggest that vascular compression of the phrenic nerve in the neck may occur following iatrogenic or terrible injuries, and lead to symptomatic diaphragm paralysis.**

Keywords: **Red Cross Syndrome, diaphragm paralysis, cervical motor nerve cells.**

1. INTRODUCTION

Although diaphragm paralysis is a rather uncommon issue, the respiratory disruptions that it causes can be crippling for those unfortunate patients in whom it happens. Generally, clinicians have recommended to afflicted patients they learn to cope with the condition, anticipating spontaneous improvement and acknowledging limited treatment choices. Typical presenting symptoms in patients with diaphragm paralysis consist of: shortness of breath with exertion and/or when supine, increased tiredness, loss of energy, gastrointestinal reflux and bloating (left-sided paralysis), and sleep disturbances.

There are several etiologies that have been described, most relating to surgical, anesthetic, or chiropractic complications in the neck and/or chest [1-8]. Terrible events that trigger a traction-type injury when the neck is jolted in an opposite direction from the shoulder and arm has been implicated as a reason for diaphragm paralysis, although is more commonly connected with injury to the brachial plexus [9]. There are generalized neuromuscular disorders, such as amyotrophic lateral sclerosis, Pompe disease, and diabetic peripheral neuropathy, which might provide with, or include diaphragm paralysis as part of their sequelae [10-12] Frequently, nevertheless no clear etiology exists, and patients will be identified as having a variation of Parsonage-Turner syndrome, or categorized with an idiopathic diaphragm paralysis [13,14]

Until recently, there has actually been no way to reverse the paralysis. Rather, some patients are used a diaphragm plication to increase lung volumes by flattening the diaphragm in its inspiratory position. Whereas this might be an effective method for reducing respiratory deficits, restoration of spontaneous diaphragmatic activity remains the supreme objective. Phrenic nerve surgery has actually recently been shown as an effective way to reverse diaphragm paralysis, and in many cases, has likewise offered insight into the underlying pathophysiology of the paralysis condition [15].

We provide 3 cases of symptomatic diaphragm paralysis that were figured out to be from vascular compression of the phrenic nerve following iatrogenic or distressing injuries.

2. METHODOLOGY

We retrospectively reviewed 3 patients who provided with persistent, symptomatic unilateral diaphragm paralysis that failed to improve with conservative management over a minimum 6 month period. They were picked from a larger accomplice of patients with diaphragm paralysis, based upon favorable findings of a vascular compression of the phrenic nerve. Two of the patients had actually experienced a preceding traction injury, one from a fall on an outstretched arm and the other from a sleep-related positional injury. The 3rd patient had actually undergone an interscalene nerve block for shoulder surgery. The Institutional Review Board at our health center approved the research study and notified approval was obtained in accordance with study approval.

In all patients diaphragm paralysis was verified on fluoroscopic chest radiographs (SNIFF), revealing either lack of diaphragmatic activity on the involved side, or paradoxical motion. Lung spirometry (PFT) was carried out to record a restrictive pulmonary deficit consistent with diaphragm paralysis. All patients underwent nerve conduction screening (NCS) of the phrenic nerve and electromyography (EMG) of the diaphragm to verify the presence of a compression neuropathy of the phrenic nerve with conservation of the motor end-plates. Extra radiographic imaging studies were acquired (MRI, CT) to rule out the possibility of degenerative cervical disc disease, or a mass in the neck, chest, or mediastinum cavity.

When no spontaneous improvement was kept in mind scientifically or radiographically, surgery was offered after a minimum of 6 months from the beginning of symptoms. All patients underwent exploration and decompression of the phrenic nerve, and nerve screening was performed intra-operatively both in the past, and after decompression to assist in predicting clinical improvement. Vascular compression of the phrenic nerve was treated by ligation of the transverse cervical artery (TCA). The adherence between the nerve and the artery was launched and a microscopic neurolysis was performed to eliminate the fibrous tissue from the compressed portion of the phrenic nerve. An anti-inflammatory agent (triamcinolone 40 mg/mL) was then penetrated into the injury cavity to prevent postoperative scar tissue and fibrosis.

All 3 patients were discharged from the medical facility on post- operative the first day. A program of pulmonary rehabilitation was initiated at 3 weeks, and continued for approximately four months postoperatively to make the most of early improvements. Follow-up examination consisted of SNIFF screening and lung spirometry at 3 months, and subjective reports of enhancements in respiratory function.

3. RESULTS AND DISCUSSION

There were 2 males and one woman, with an average age of 49 (range 40-54 yrs) [see Table 1] In two patients the diaphragm paralysis was left-sided, whereas a right-sided diaphragm paralysis existed in one patient. In 2 patients the diaphragm paralysis had existed for greater than one year without enhancement, whereas the diagnosis had been made eight months prior in one patient.

All 3 patients explained shortness of breath symptoms exacerbated with exertion. In one patient abrupt, severe dyspnea took place when turning her head to the affected side. Sleep disturbances were reported by all patients, two of whom needed nighttime CPAP for considerable sleep-disordered breathing. In the patients with left-sided diaphragm paralysis, symptoms of bloating and GERD were reported. One patient was on complete medical disability due to the respiratory symptoms of diaphragm paralysis and a second patient was on restricted duty, unable to perform manual work. The third patient was an executive who might perform daily work obligations.

All patients had SNIFF tests showing an unequivocal unilateral diaphragm paralysis and spirometry results consistent with a mild-to-moderate restrictive ventilatory deficit. The results of NCS/EMG testing exposed the following conduction speeds (imply 13.53 ms, variety 7.7- 16.8 ms; [ref. 8.0 ± 1.5 ms] and motor unit possible amplitudes (indicate 0.21 mV, range 0.02-- 0.4 mV; [ref. ≥ 0.33 mV] [see Table 1] PFT screening results in all patients revealed a limiting ventilatory deficit in the mild-to-severe range (mean FEV1 66%, range 60-74%; imply FVC 67%, range 52-76%). Of note, the patient whose head turning exacerbated dyspneic symptoms showed a conduction speed of 7.7 ms and motor amplitude of 0.4 mV with her head in a neutral position (typical values), however no action was detectable in either test with her head turned to the.

The surgical procedure performed in all 3 patients uncovered a vascular compression of the phrenic nerve in the neck, specifically at the area where the TCA crosses above it (see Fig. 1). In one patient the TCA appeared dilated and tortuous, whereas in the other two patients there was an apparent thick adherence in between the nerve and the vessel, constant with

fibrosis. Intra-operative nerve screening prior to decompression corroborated pre-operative findings. In all 3 patients attempts to promote the phrenic nerve above the site of vascular compression failed to generate a diaphragmatic response, whereas a response was recorded with stimulation distal to the compression. Following nerve decompression, stimulation of the phrenic nerve all along its course in the neck resulted in a diaphragmatic response at physio- reasoning thresholds (0.5-1.0 mA) (see Fig. 2). All patients provided subjective reports of enhancements in their breathing function within the very first 48 h after surgery. Specif- ically, they noticed having the ability to take much deeper breaths, and were able to lie supine without an exacerbation of symptoms. The one patient whose dyspnea manifested with a head turn, was now able to per- kind that steer without such symptoms occurring. At three months SNIFF tests and PFTs were carried out, revealing motion of the formerly paralyzed diaphragms and a normalization of the restrictive spirometry deficits, respectively. At 6 months, all patients reported an upkeep or improvement of early outcomes, and they were all able to return to regular work duties. The patients needing nighttime CPAP had the ability to stop this therapy in the very first 3 months after surgery. There were no late or early surgical complications.

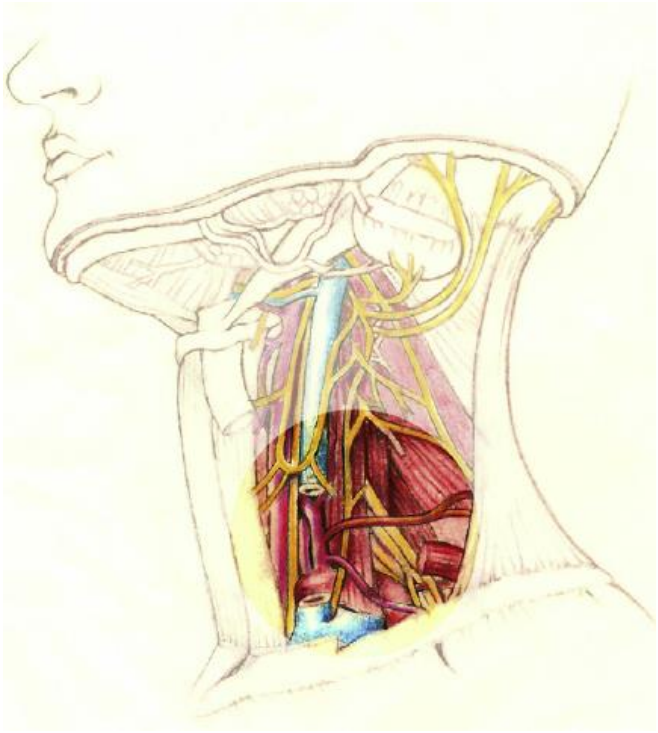

Fig. 1. The transverse cervical artery (TCA) is one of the branches arising from the thyrocervical trunk and coursing laterally in the lower neck. The TCA intersects the phrenic nerve at an almost 90◦ angle, and following trauma or iatrogenic injury there can be adherence between the two structures, resulting in a vascular compression neuropathy.

Peripheral reasons for diaphragm paralysis are frequently due to a phrenic nerve injury in the chest, neck, or mediastinum, whereas spine injury, brain growth, and central hypoventilation syndrome are the most likely central nerve system etiologies. Understanding the pathophysiology of phrenic nerve injury has been hypothesized, but rarely validated, because the majority of affected patients are handled non-operatively. Even in patients who have actually been treated with diaphragm plication there is normally no exposure or expedition at the site of nerve injury, hence it is unusual for the underlying injury to be straight examined. There are couple of reports of immediate phrenic nerve repair work in patients undergoing growth ablation, or following chest injury, where paperwork of nerve transection has actually occurred [16,17].

Table 1 Demographics and outcomes of patients with vascular compression of the phrenic nerve

Patient	Age	Sex	Side	NCS (ms)	EMG (mV)	FEV1	FVC	Recovery
1	40	M	L	16.8	0.02	74%	76%	+
2	52	F	R	7.7/NDa	0.4/NDa	60%	52%	+
3	54	M	L	16.09	0.2	62%	73%	+

ND – patient 2 had no detectable NCS/EMG response with her head turned to the right .

Our recent publication examined a small series of patients with diaphragm paralysis treated with nerve surgery techniques; documenting successful diaphragm re-innervation in 89% [15] Expedition at the website of nerve injury supplied an opportunity to better comprehend the pathophysiology of nerve injury in a few of these cases. Peripheral nerve injuries including the nerves of the upper and lower extremities have been thoroughly investigated and, contrary to phrenic nerve injuries, are much better comprehended. Despite the system of injury (i.e. transection, extending, thermal, crush, compression), the outcome will be damage to one or more of the supporting nerve structures and/or axons that, if extreme enough, will result in Wallerian degeneration the atrophy of the nerve structure distal to the site of injury. There are comprehensive nerve injury classifications by Seddon and Sunderland that help us comprehend the seriousness of the nerve injury, and also to prognosticate the opportunities of spontaneous healing [18,19]

Post-procedural and post-surgical nerve injuries may be from direct nerve transection or extending, or may be secondary to the fibrosis and scarring that occurs after such occasions. Loss of typical tissue aircrafts and adherence between generally separate anatomical structures can result in compression and nerve dysfunction. When it comes to the phrenic nerve, its course in the neck, running in-line with the anterior scalene muscle and just under the prevertebral fascia, might predispose it to compression injuries following neck interventions. We think that injury or adjustment in the neck might result in scalene muscle induration, sometimes in association with an intramuscular hematoma, causing compression of the phrenic nerve where it is interposed between the muscle and the semi-rigid prevertebral fascia.

The TCA, a branch of the thyrocervical trunk developing from the subclavian artery, crosses the phrenic nerve around 3 cm above the clavicle, nevertheless is anatomically different from the nerve by the distinct prevertebral fascia. Following intervention in the neck, it is very imaginable that a change or disturbance in the regular quality of the prevertebral fascia might result in adherence between the two structures. The course and caliber of the TCA might also be changed in a way that triggers greater compression of the underlying phrenic nerve. Although a vascular nerve compression may lead to either a demyelinating or ischemic neurapraxia, early reversibility of the paralysis following treatment in our three cases supports the former as the underlying pathologic mechanism.

Vascular compression of a central or peripheral nerve has actually been well explained in various locations throughout the body. Test- ples of such syndromes include: vertebral artery compression of a cervical root, arterial compression of the intracranial trigeminal nerve, thoracic outlet syndrome, vascular compression of the vestibulocochlear nerve, radial nerve palsy secondary to the vascular leash of Henry, Ortner's syndrome (vascular compression of the frequent laryngeal nerve), and vascular compression of the occipital nerve causing migraine headaches [20-25] .In particular, radial nerve palsy secondary to a thickened reoccurring radial artery (vascular leash of Henry) may take place in a comparable way to our suggested syndrome because, preceding trauma or inflammation changes the caliber or course of the capillary and/or spatial relationship in between it and the included nerve.

Regrettably, it is unlikely that even the most astute clinician would have the ability to definitively identify vascular compression to be the reason for a patient's diaphragm paralysis. Maybe a high index of suspicion for such a syndrome could be generated in a patient whose dyspnea symptoms are exacerbated with head turning, however there is no way to support this with radiographic findings. Unlike thoracic outlet syndrome, in which a contrast-enhanced MRI may often visualize vascular compression of the brachial plexus in association with a positive Adson's maneuver, the TCA and phrenic nerve are not well envisioned, even with the most advanced of imaging research studies. "Red Cross syndrome" may precisely portray the pictured course of the TCA over the phrenic nerve at a practically perfect 90 ◦ angle; nevertheless this diagnosis is one that will often be made intra-operatively until there are substantial advances in our capability to image little caliber nervous and vascular structures. Transcutaneous stimulation of the phrenic nerve with a nerve conduction probe, in conjunction with chest fluoroscopy, can determine a delay in conduction velocity that supports a diagnosis of compression neuropathy, however the area and source of compression frequently cannot be precisely determined.

Clinicians examining patients with symptomatic diaphragm paralysis need to be conscious that there are reversible conditions such as vascular compression, and patients ought to be notified that treatments for these conditions do exist. A reversible compression neuropathy of the phrenic nerve is a diagnosis suggested by particular findings on phrenic nerve NCS and diaphragm EMG, along with a helpful clinical history. Vascular compression is just one of the mechanisms by which the phrenic nerve can be reversibly, or irreversibly, compressed, yet peripheral nerve surgeons have reputable surgical techniques for handling whatever condition emerges as the underlying patho-physiology.

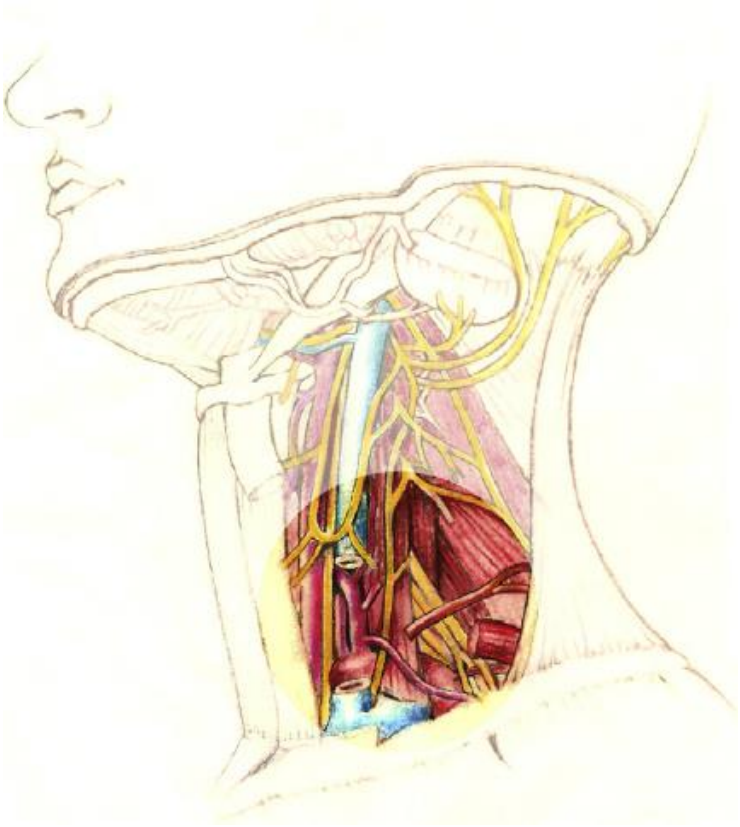

Fig. 2. Ligation of the TCA, along with a more extensive micro-neurolysis of the phrenic nerve, may reverse an ischemic neurapraxia and restore diaphragmatic function in patients with paralysis.

4. CONCLUSION

Little is learnt about the pathophysiology of phrenic nerve injuries, especially given that treatment options have generally been restricted. Current advances in surgical treatment have actually enabled us to begin to recognize underlying pathophysiology. We present three patients whose diaphragm paralysis was eventually triggered by a reversible vascular compression from an adherent or tortuous TCA. Increased awareness of conditions, such as Red Cross syndrome, will permit clinicians to much better comprehend the causes of diaphragm paralysis, and hopefully lead to greater numbers of patients finding effective treatments for this disorder.

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