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Diaphragm paralysis caused by transverse cervical artery compression of the phrenic nerve: The Red Cross syndrome

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ABSTRACT

Background: The etiology of diaphragm paralysis is often elusive unless an iatrogenic or traumatic injury to the phrenic nerve can be clearly implicated. Until recently, there has been little interest in the pathophysiology of diaphragm paralysis since few treatment options existed.
Methods: We present three cases of symptomatic diaphragm paralysis in which a clear clinico-pathologic diagnosis could be identified, specifically a vascular compression of the phrenic nerve in the neck caused by a tortuous or adherent transverse cervical artery.
Results: In two patients the vascular compression followed a preceding traction injury, whereas in one patient an inter-scalene nerve block had been performed. Following vascular decompression, all three patients regained diaphragmatic motion on fluoroscopic chest radiographs, and experienced a resolution of respiratory symptoms.
Conclusion: We suggest that vascular compression of the phrenic nerve in the neck may occur following traumatic or iatrogenic injuries, and result in symptomatic diaphragm paralysis.

1. Introduction

Although diaphragm paralysis is a rather uncommon problem, the respiratory disturbances that it causes can be debilitating for those unfortunate patients in whom it occurs. Traditionally, clinicians have suggested to affected patients they learn to live with the condition, anticipating spontaneous improvement and acknowledging limited treatment options. Common presenting symptoms in patients with diaphragm paralysis include: shortness of breath with exertion and/or when supine, increased fatigue, 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]. Traumatic events that cause a traction-type injury – when the neck is jolted in an opposite direction from the shoulder and arm – has been implicated as a cause of diaphragm paralysis, although is more commonly associated with injury to the brachial plexus [9]. There are generalized neuromuscular disorders, such as amyotrophic lateral sclerosis, Pompe disease, and diabetic peripheral neuropathy, which may present with, or include diaphragm paralysis as part of their sequelae [10–12]. Often, however no clear etiology exists, and patients will be labeled as having a variant of Parsonage-Turner syndrome, or classified with an idiopathic diaphragm paralysis [13,14].

Until recently, there has been no way to reverse the paralysis. Instead, some patients are offered a diaphragm plication to increase lung volumes by flattening the diaphragm in its inspiratory position. Whereas this may be an effective method for reducing respiratory deficits, restoration of spontaneous diaphragmatic activity remains the ultimate goal. Phrenic nerve surgery has recently been demonstrated as an effective way to reverse diaphragm paralysis, and in some cases, has also provided insight into the underlying pathophysiology of the paralysis condition [15].

We present three cases of symptomatic diaphragm paralysis that were determined to be from vascular compression of the phrenic nerve following traumatic or iatrogenic injuries.

2. Methods

We retrospectively reviewed three patients who presented with chronic, symptomatic unilateral diaphragm paralysis that failed to improve with conservative management over a minimum six month period. They were selected from a larger cohort of patients

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with diaphragm paralysis, based upon positive findings of a vascular compression of the phrenic nerve. Two of the patients had experienced a preceding traction injury, one from a fall on an outstretched arm and the other from a sleep-related positional injury. The third patient had undergone an inter-scalene nerve block for shoulder surgery. The Institutional Review Board at our hospital approved the study and informed consent was obtained in accordance with study approval.

In all patients diaphragm paralysis was confirmed on fluoroscopic chest radiographs (SNIFF), revealing either absence of diaphragmatic activity on the involved side, or paradoxical motion. Lung spirometry (PFT) was performed to document a restrictive pulmonary deficit consistent with diaphragm paralysis. All patients underwent nerve conduction testing (NCS) of the phrenic nerve and electromyography (EMG) of the diaphragm to confirm the existence of a compression neuropathy of the phrenic nerve with preservation of the motor end-plates. Additional radiographic imaging studies were obtained (MRI, CT) to rule out the possibility of degenerative cervical disc disease, or a mass in the neck, mediastinum, or chest cavity.

Surgery was offered after a minimum of six months from the onset of symptoms when no spontaneous improvement was noted clinically or radiographically. All patients underwent exploration and decompression of the phrenic nerve, and nerve testing was performed intra-operatively both before, 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 artery and the nerve was released and a microscopic neurolysis was performed to remove the fibrous tissue from the compressed portion of the phrenic nerve. An anti-inflammatory agent (triamcinolone 40 mg/mL) was then infiltrated into the wound cavity to prevent post-operative scar tissue and fibrosis.

All three patients were discharged from the hospital on post-operative day one. A program of pulmonary rehabilitation was initiated at three weeks, and continued for up to four months post-operatively to maximize early improvements. Follow-up evaluation included SNIFF testing and lung spirometry at three months, and subjective reports of improvements in respiratory function.

3. Results

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

All three patients described shortness of breath symptoms exacerbated with exertion. In one patient sudden, severe dyspnea occurred when turning her head to the affected side. Sleep disturbances were reported by all patients, two of whom required nocturnal CPAP for significant sleep-disordered breathing. In the patients with left-sided diaphragm paralysis, symptoms of bloating and GERD were reported. One patient was on full medical disability due to the respiratory symptoms of diaphragm paralysis and a second patient was on restricted duty, unable to perform manual labor. The third patient was an executive who could perform daily work responsibilities.

All patients had SNIFF tests demonstrating an unequivocal unilateral diaphragm paralysis and spirometry results consistent with a mild-to-moderate restrictive ventilatory deficit. The results of NCS/EMG testing revealed the following conduction velocities (mean 13.53 ms, range 7.7–16.8 ms; [ref. 8.0 ± 1.5 ms]) and motor unit potential amplitudes (mean 0.21 mV, range 0.02–0.4 mV;[ref. $\geq$0.33 mV]) [see Table 1]. PFT testing results in all patients revealed a restrictive ventilatory deficit in the mild-to-severe range (mean FEV1 66%, range 60–74%; mean FVC 67%, range 52–76%). Of note, the patient whose head turning exacerbated dyspneic symptoms demonstrated a conduction velocity of 7.7 ms and motor amplitude of 0.4 mV with her head in a neutral position (normal values), however no response was detectable in either test with her head turned to the right.

The surgical procedure performed in all three patients uncovered a vascular compression of the phrenic nerve in the neck, specifically at the location where the TCA crosses above it (see Fig. 1). In one patient the TCA appeared tortuous and dilated, whereas in the other two patients there was an obvious dense adherence between the vessel and the nerve, consistent with fibrosis. Intra-operative nerve testing prior to decompression corroborated pre-operative findings. In all three patients attempts to stimulate the phrenic nerve above the site of vascular compression failed to elicit 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 physiologic thresholds (0.5–1.0 mA) (see Fig. 2).

All patients provided subjective reports of improvements in their respiratory function within the first 48 h after surgery. Specifically, they noticed being able to take deeper breaths, and were able

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/ND[a]	0.4/ND[a]	60%	52%	+
3	54	M	L	16.09	0.2	62%	73%	+

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

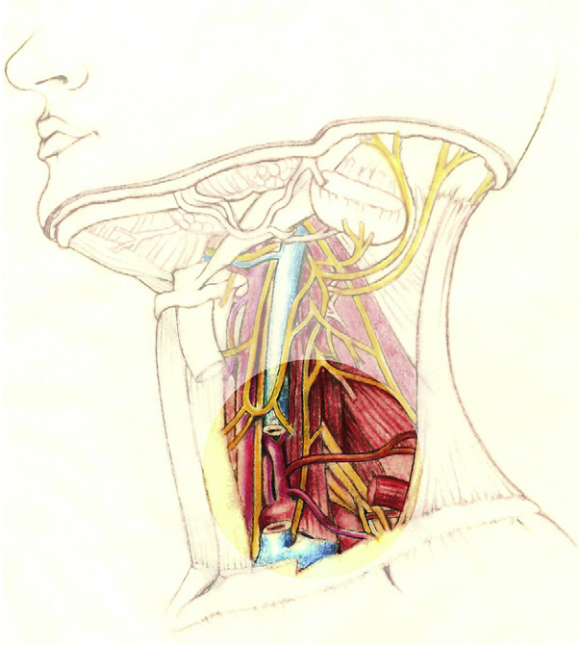

Fig. 1. The transverse cervical artery (TCA) is one of the branches arising from the thyrocervical trunk and coursing laterally in the lower neck. The TCA intersects the phrenic nerve at an almost 90° angle, and following trauma or iatrogenic injury there can be adherence between the two structures,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