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CLINICAL CASE REPORT

Phrenic nerve neurotization using the spinal accessory nerve for diaphragmatic palsy in extensive high spinal cord injury secondary to idiopathic acute transverse myelitis

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Abstract

The authors present a case of functional improvement of diaphragmatic paralysis in extensive high spinal cord injury, performing a neurotization of the phrenic nerve with the spinal accessory nerve. A C2-C5 injury of the spinal cord was diagnosed in a 15 years old female, secondary to idiopathic acute transverse myelitis. The patient did not have automatic respiration at the time of mechanical ventilation removal; moreover, she presented ventilatory distress. The neurotization of the left phrenic nerve with left spinal accessory nerve was performed 3 months after the injury, at six months after surgery she showed mobilization of the left hemidiaphragm and reached mechanical ventilation independence.

Key words: Phrenic nerve, Spinal accessory nerve, Neurotization, Diaphragmatic palsy

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Introduction

In spinal cord injury, when the phrenic nerve nucleus is affected, it can produce diaphragmatic muscle paralysis that affects the respiratory pattern, being necessary the use of mechanical ventilation for long periods. This is associated with degeneration of nerve fibers, resulting in damage of the nuclear cells of the phrenic nerve and diaphragm weakness, because a myofibrillar and myofilament protein loss occurs causing diaphragmatic dysfunction [1]. Although, the spinal accessory nerve (SAN) have a supralesional origin and remains intact, innervating the sternocleidomastoid and trapezius muscles, also participates in the breathing process helping in inspiration. The nerve transfers have shown a successful reactivation of injured nerves and some authors have proposed nerve transfers for diaphragmatic dysfunction [2, 3].

We present a case in which neurotization of the phrenic nerve was performed with the SAN, allowing re-innervate the diaphragmatic muscle, improving respiratory function.

Case Report

A 15-year-old female who starts suddenly without making any effort, with loss of strength in the lower limbs, progressing to the upper limbs and with respiratory distress, bringing her to our hospital, where upon admission she presented cardiopulmonary arrest demanding advanced cardiopulmonary resuscitation during a two minutes cycle and required endotracheal intubation and mechanical ventilatory support. Subsequently she was

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admitted to the ICU and 3 days after, a MRI of the brain and spine was performed showing a C2 to C5 diffuse spinal cord lesion. Five days after admission, the sedation was retired, finding a conscious, awake patient, with no strength in the four extremities 0/5, and a sensory level affected at C2 and distal, absence of muscle stretch reflexes, no control of the sphincters and without respiratory effort. The diagnosis of idiopathic acute transverse myelitis was realized once neuromyelitis optica and multiple sclerosis were ruled out.

At 3 weeks the tracheostomy and gastrostomy were performed, multiple attempts were made to remove mechanical ventilation and nevertheless the patient did not present spontaneous ventilation. The attempts to withdraw mechanical ventilation continued for 3 months, making it impossible to achieve independent from mechanical ventilation. A lung fluoroscopy is performed showing weak right hemidiaphragm movement, but total palsy of the left hemidiaphragm. Transfer of the XI to the left phrenic nerve was proposed to improve breathing.

Operation

The patient was positioned supine with the head turned to the right. In the left supraclavicular space an inverted "L" incision was made above the clavicle and on the posterior border of the sternocleidomastoid muscle. The SAN was located with electrical stimulation; we proceed to locate the phrenic nerve in the same way. The SAN was transected and moved 5 cm under the sternocleidomastoid muscle and was coapted with the sectioned phrenic nerve applying fibrin glue, giving us a direct free tension termino-terminal anastomosis, without the use of nerve graft (Figure 1).

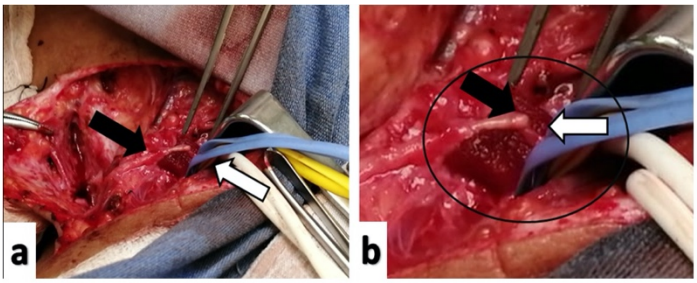

Figure 1 - -terminal anastomosis of the phrenic nerve (black arrow) and the spinal accessory nerve (white arrow). a A tensionless anastomosis. b Enlarged view of the anastomosis that is inside the circle.

Evolution

Six months after the surgery, the patient presented left diaphragmatic mobilization, as well as respiratory automatism and total mechanical ventilatory independence. She is currently 1 year without mechanical ventilation or supplemental oxygen and mobilizes the upper limbs with strength 4/5 and the lower limbs 2/5. However sphincter dysfunction persists and the patient needs a wheelchair for her displacement (Figure 2).

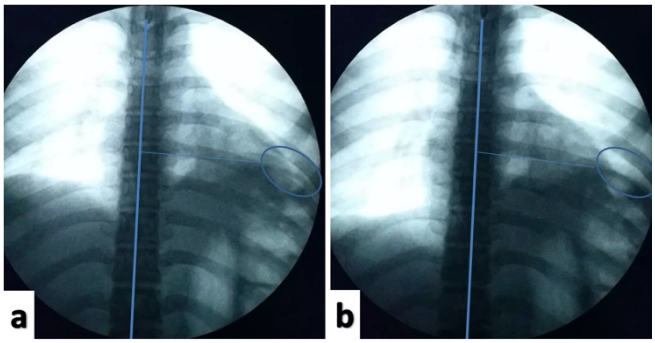

Figure 2 - Chest Fluoroscopy showing left diaphragm movement. a Exhalation. b Inhalation.

Discussion

The respiratory dysfunction has been associated with immune-mediated demyelinating diseases, although is rare and trends to spontaneous partial or sometimes complete recovery, when occurs is catastrophic for the patient, resulting in lifelong mechanic ventilatory support and sometimes the death. [4, 5]

The SAN originate from the spinal nucleus of the lateral gray matter of the cervical spinal levels C1 to C5 and 4-5 rootlets originated from the dorsolateral surface of the medulla oblongata. It has 1700 - 2000 nerve fibers and 2 mm width [2].

The phrenic nerve originates from the motor nucleus of the phrenic located in the medial part of the gray matter of the C3-C5 medullary segments. It has 3500 nerve fibers and is 2.5 mm width [2].

Albeit the accessory phrenic nerve must be kept in mind at the time of surgery, it has his origin in C3-C5 medullar segments and would not be expected to function in innervating the diaphragm muscle spontaneously [6].

Gordon T [7] has shown the rate of nerve regeneration after injury is 1 mm/d their findings are based on adults. Ladak A [8] found elbow flexion at 6 months after nerve transfers in obstetrical brachial plexus injuries.

The mechanism by which the SAN can replace the function of the phrenic nerve, follows the principles of nerve transfers, in which a nerve with a different function can perform other functions than the original ones by reinnervating a nerve that have lost its function, as has been demonstrated with the different nerve transfers [9].

Kaufman MR [10] in two adult patients with C1-C7 lesion performed a nerve transfer with the complete SAN to the phrenic nerve without a diaphragm pacemaker reaching ventilator independence at 12 and 24 months after the surgery.

Nandra KS [11] have performed end-to-end anastomosis of the intercostal nerves to the phrenic nerve in four adults patients with C3-C5 spinal cord

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injury; additionally they placed diaphragmatic pacing, although they achieve a significant reduction in dependence on mechanical ventilation .

In our previous pediatric patient with diaphragmatic palsy secondary to high spinal cord injury we use the half of SAN and reinnervating the diaphragm muscle, with a termino-lateral anastomosis improving the respiratory function, although did not reach total ventilatory independence, the entire SAN could have been a better choice [12].

Yang M [13] in their reactivation of the diaphragm used the entire SAN transfer to the phrenic nerve with a termino-lateral anastomosis, achieving ventilatory independence one year after surgery in an adult patient with a post traumatic C2 injury; we have the same results at 6 months after surgery with the use of entire SAN and a termino-terminal anastomosis.

Lareille J [14] in a 5 year old female with a lesion at C1 and tetraplegia performed an intercostal to phrenic nerve transfer and applied a phrenic nerve pacing, achieving ventilatory independence 2 years after surgery.

Verin E [15] used the inferior laryngeal nerve to phrenic nerve transfer in four adult patients with C3-C5 injury and applied a diaphragmatic pacing; nevertheless no one patient reached automatic ventilation.

Conclusion

Results can be attributed to increase potential for nerve regeneration and better cortical plasticity in infants. Total mechanical ventilation independence was achieved, could be associated to the use of entire SAN to re-innervate the phrenic nerve.

This surgical technique could be considered an option to avoid prolonged periods of mechanical ventilation in patients with high spinal cord injury and reach ventilatory autonomy.

To our knowledge is the first case report showing de use of complete SAN to the phrenic nerve transfer and termino- terminal anastomosis in one pediatric patient with successful surgery.

Disclosure Statement

The authors have no conflicts of interest to declare.

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