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CLINICAL INFORMATION

Supraclavicular nerve and superior trunk block for surgical treatment of clavicle fracture in a patient with Steinert's disease – Case report☆

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Abstract
Background and objectives: Ultrasound-guided upper limb blocks may provide great benefits to patients with serious diseases. Patients with Steinert's disease have muscle weakness and risk of triggering myotony or malignant hyperthermia due to the use of anesthetic agents and surgical stress. The objective of this report was to demonstrate a viable alternative for clavicle fracture surgery with upper trunk and supraclavicular nerve block, thus reducing the spread of local anesthetic to the phrenic nerve in a patient with muscular dystrophy.
Case report: A 53-year-old male patient with Steinert's disease, associated with dyspnea, hoarseness and dysphagia, referred to the surgical theater for osteosynthesis of clavicle fracture. Upper limb (1 mL 0.75% ropivacaine) and supraclavicular nerve block (1 mL 0.75% ropivacaine in each branch) were combined with venous anesthesia with propofol under laryngeal mask (infusion pump target of 4 mcg.mL^{-1}). Upon awakening, the patient had no pain or respiratory complaints. He was transferred to the ICU for immediate postoperative follow-up with discharge from this unit after 24 h without complications.
Conclusions: The superior trunk and cervical plexus block associated with venous anesthesia under laryngeal mask, without the use of opioids, proved to be adequate in the case of a patient with clavicle fracture and Steinert's disease.

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With the use of ultrasonography in regional anesthesia it is possible to perform increasingly selective blocks, thus allowing greater security for the anesthetic-surgical procedure and lower morbidity for the patient.

PALAVRAS-CHAVE
Doença de Steinert;
Distrofia muscular;
Complicações pós-operatórias;
Clavícula;
Nervos periféricos;
Ultrassom

Bloqueio do nervo supraclavicular e do tronco superior para tratamento cirúrgico de fratura de clavícula em paciente portador de doença de Steinert – Relato de caso

Resumo
Justificativa e objetivos: Bloqueios seletivos dos membros superiores guiados por ultrassom podem trazer grandes benefícios em pacientes portadores de doenças graves. Pacientes portadores da doença de Steinert apresentam fraqueza muscular e riscos de desencadear miotonia ou hipertermia maligna devido ao uso de agentes anestésicos e ao estresse cirúrgico. O objetivo deste relato foi mostrar uma opção viável para a cirurgia de fratura de clavícula com bloqueio do tronco superior e nervo supraclavicular, diminui-se assim a dispersão do anestésico local para o nervo frênico em paciente com distrofia muscular.
Relato de caso: Paciente do sexo masculino, 53 anos, portador de doença de Steinert, associada a dispneia, rouquidão e disfagia. Encaminhado ao bloco cirúrgico para osteossíntese de fratura de clavícula. Feito bloqueio de tronco superior (1 mL ropivacaína a 0,75%) e de nervo supraclavicular (1 mL de ropivacaína 0,75 em cada ramificação) associado à anestesia venosa com propofol sob máscara laríngea (alvo de 4 $mcg.mL^{-1}$ em bomba de infusão). Ao despertar, o paciente apresentava-se sem dor ou queixas respiratórias. Admitido em CTI para acompanhamento do pós-operatório imediato com alta dessa unidade após 24 horas sem intercorrências.
Conclusões: O bloqueio do tronco superior e do plexo cervical associado à anestesia venosa sob máscara laríngea, sem uso de opioides, mostrou-se adequado no caso de fratura da clavícula em paciente com doença de Steinert. Com o uso da ultrassonografia em anestesia regional é possível fazer bloqueios cada vez mais seletivos e possibilitar assim maior segurança para o procedimento anestésico-cirúrgico e menor morbidade para o paciente.

Introduction

Described by Steinert (Germany, 1909), Steinert's disease is the most common form of muscular dystrophy in adults. Its prevalence is estimated between 3 and 5 cases per 100,000 inhabitants, with incidence of one case for every 8000 births. It is genetically transmitted in an autosomal dominant way and in most cases the gene is inherited from the mother. The genetic mutation characteristic of Steinert's disease is a repeated sequence of the cytosine, thymine, and guanine nitrogen bases of variable length on chromosome 19. The extension of this sequence determines the clinical involvement of the disease. In general, the disease manifestation occurs between the second and fourth decades of life, marked by hand paresis, weakness of foot dorsiflexion, cataract, and infertility. The characteristic facial appearance of patients is due to muscular weakness and atrophy of the face and cervical region.

Steinert's disease is a myotonic dystrophy and its main characteristic is the presence of myotonia, which is the persistence of active skeletal muscle contraction even after the initial voluntary effort has ceased. Atrophy and muscle weakness progressively occur. It is emphasized that myotonia is an intrinsic disorder of the muscle, not the peripheral nerve or neuromuscular junction. Thus, myotonia is not abolished by peripheral nerve block or neuromuscular blocker administration. It is believed that the persistence of muscle contraction is caused by injury to the chlorine or sodium channels of the muscle membrane, which decreases chlorine conductance and, in turn, results in prolonged contracture.

Steinert's myotonic dystrophy is considered a multisystemic disease, since, in addition to muscular involvement, there is interference in the functioning of other systems and organs, may cause cataracts, cardiac conduction system deterioration, tachyarrhythmia, cardiomyopathy, valvular disease, pulmonary disease (due to reduced gastric emptying time), gastroesophageal reflux, cholelithiasis, constipation, intestinal pseudo-obstruction, cognitive dysfunction, mental retardation, frontal baldness, hypothyroidism, hypogonadism, dyspnea, infertility, growth hormone secretion disorders, abnormal glucose and insulin metabolism. Due to pulmonary involvement, patients may present with respiratory muscle weakness and disturbances in central control of breathing. Reduced ventilatory response to carbon dioxide may occur, and chronic hypercarbia is a common condition in these patients. Bisinotto et al. also reported a higher probability of malignant hyperthermia in patients with Steiner's disease.[1]

The potential triggers of myotonia are fear; prolonged fasting; hypokalemia; hypoxemia; hypercarbia; tourniquet; pain; anxiety; adrenergic discharge; electric scalpel; hypothermia; peripheral nerve stimulator; tremors; voluntary effort; electrical or mechanical stimulation before, during, or after surgery; drugs (e.g. propranolol and potassium); and anesthetic agents, such as succinylcholine and anticholinesterases.

The most common complications in these patients are pulmonary such as acute ventilatory failure, but the main causes of mortality are cardiovascular complications, such as ventricular dysfunction, myocardial ischemia, pulmonary embolism, ventricular tachycardia, asystole, and pulseless electrical activity that result in sudden death.

Clavicle fractures represent 35% of the scapular girdle lesions and occur more frequently in men. They are commonly caused by falls during cycling and equestrian sports. The peripheral nerve block technique best suited to anesthetize the clavicle is still controversial. Different strategies are described in the literature, such as superficial cervical plexus block, combined superficial and deep block, and interscalene nerve block.

There is a consensus in the literature that analgesia on the skin over the clavicle is innervated by the supraclavicular nerve. However, bone inversion is controversial.[2] Authors correlate the contribution of both the cervical plexus and the brachial plexus with their different ramifications (subclavian nerve, suprascapular nerve, and long thoracic nerve) to the bony innervation of the clavicle.[2] Based on this, the association of brachial plexus and cervical plexus would still be the most appropriate for anesthesia in cases of clavicle fractures.

The cervical plexus block encompasses the supraclavicular nerve. However, with this technique the entire cervical plexus, including the phrenic nerve, is blocked. The supraclavicular nerve emerges from the cervical plexus, following a caudal, superficial pathway to the middle scalene muscle, differently from the phrenic nerve, which follows the caudal course, but on the anterior scalene muscle. With the use of ultrasound it is possible to block the supraclavicular nerve with small volumes of local anesthetic and at lower levels, reducing the possibility of phrenic nerve paralysis.[2]

The classic interscalene technique is performed at the cricoid cartilage level (C6), where it is located in the sulcus between the anterior and middle scalene muscles with a high index of phrenic nerve block. With the advent of ultrasound, more selective blocks of the C5 and C6 roots (just below the transverse process of C6) have been used, with small volumes of local anesthetic. However, this technique is still associated with risks of phrenic nerve palsy, with dorsal scapular nerve and long thoracic nerve injury, as well as postoperative neurological symptoms.[3] Below the transverse process of C7, the C5 and C6 roots become increasingly superficial, progressively more distant from the phrenic nerve, and bind to form the superior trunk, which is visible as the only complex structure interspersed with hyperechogenic connective tissue, which is found just below the prevertebral fascia.[3]

The improved anatomical knowledge provided by ultrasonography enabled anesthesiologists to design new locoregional blocking techniques and refine the existing ones. The upper trunk and supraclavicular nerve block is an example of this, and may be a safer option in relation to interscalene and cervical plexus block.

The present study aims to present the case of a patient with Steinert's disease undergoing surgical repair of right clavicular fracture under regional block and total intravenous anesthesia, as well as to expose the particularity, limitation, and adequacy of anesthetic management in this context.

Case report

A 53-year-old male patient, emaciated (50 kg), with Steinert's disease, who reported locomotion disorder and a history of hoarseness and recurrent episodes of dyspnea and fatigue. He denied medication allergy, other comorbidities or regular use of medications; had undergone anesthesia-surgical procedure of cholecystectomy and cataract surgery uneventfully. He presented himself at emergency department with complaint of right shoulder pain. He reported a fall from height preceded by an intense episode of palpitation, nausea, and profuse sweating. He referred trauma to his right shoulder due to fall. An X-ray showed a fracture with deviation of the proximal third of the right clavicle, and a surgical osteosynthesis under regional block associated with total intravenous anesthesia was decided.

After preparing the surgical room with care to prevent hypothermia, conventional monitoring was performed with cardioscopy, pulse oximetry, and noninvasive blood pressure (NIBP), and the patient was warmed throughout the procedure with thermal blanket. Considering that the patient was placed in the beach chair position for surgery, the noninvasive arterial pressure cuff was positioned in the left arm for monitoring for cerebral perfusion. With the patient fully awake, without sedatives, skin antisepsis and local anesthesia (''anesthetic button'' with local infiltration of 40 mg lidocaine without vasoconstrictor) was performed. The right supraclavicular nerve block was guided by ultrasound according to the technique previously described by Maybin et al.[2] Using a Stimuplex® A50 needle (22G × 2″), an ultrasound with linear transducer (Sonisite®, M-Turbo model) and a peripheral nerve stimulator (Stimuplex, B. Braun®), two ramifications (hypoechoic structures) of the supraclavicular nerve were identified (Fig. 1) and 1 mL of the 0.75% ropivacaine anesthetic solution (7.5 $mg.mL^{-1}$) was infused in each branch. In combination, the upper right trunk block was performed following the technique described by Burckett et al.[3] After identifying the fusion of the C5 and C6 roots (Fig. 2), infiltration of 10 mL of 0.75% ropivacaine anesthetic solution (7.5 $mg.mL^{-1}$) was performed (Video 1; supplementary material online). Subsequently, the induction of general anesthesia was performed with intravenous target-controlled infusion of propofol (target of 4 $mcg.mL^{-1}$). A laryngeal mask (ML) number 4 was introduced and synchronized intermittent mandatory ventilation (SIMV) was applied with the following ventilatory parameters: PMAX = 20 cm H_2O; V_T = 350 mL; RR = 12; PEEP = 6 cm H_2O; Trigger = 2 $L.min^{-1}$; Flux.Insp. = 22 $L.min^{-1}$;

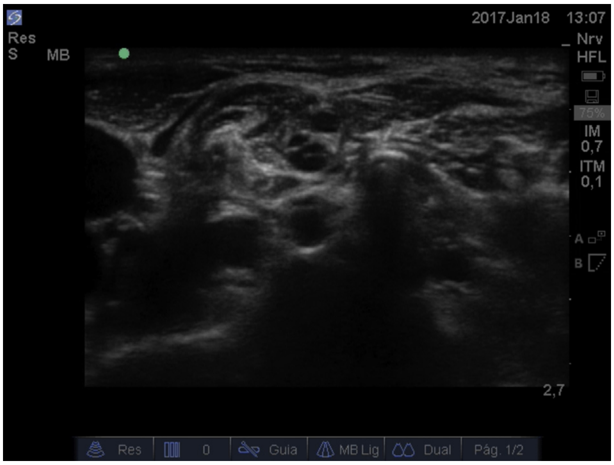

Figure 1 Ultrasound image without prominence on the left side and, on the right side, emphasizes on supraclavicular nerve branches (SCN), transverse process of C7 (TP C7), and C7, C6 and C5 roots (deep to superficial level respectively), sternocleidomastoid muscle (SCM) and anterior scalene muscle (ASM) and medium scalene muscle (MSM).

TINSP = 1.7; TIP:TI = 10%. It should be emphasized that no opioid or muscle relaxant was administered. At the end of the procedure and laryngeal mask removal, the patient was awake, conscious, lucid, alert, oriented (time and space), hemodynamically stable, eupneic (with supplemental oxygen via nasal catheter at 2 $L.min^{-1}$), with no signs of respiratory distress or discomfort, with no complaints of pain.

The patient was taken to the Intensive Care Unit (ICU) and a simple chest X-ray was performed on admission, with no evidence of phrenic nerve block. The patient had a good clinical evolution and referred only moderate pain upon surgical site manipulation after 14 h of the procedure. He was discharged from the ICU to the ward after 24 h.

Discussion

The anesthetic technique of choice for patients with Steinert's disease remains uncertain, but the use of peripheral nerve block or neuraxial block is preferred. As complications in the postoperative period usually result in pulmonary and cardiac dysfunction, general anesthesia with tracheal intubation and neuromuscular relaxation should be avoided, when possible, due to ventilatory risks inherent to postoperative control. Authors such as Cope et al. and March et al. recommend regional anesthesia in order to avoid the use of drugs potentially triggering myotonic seizures, or the combination of general-regional anesthesia with restricted use of opioids, as these patients are at high risk for respiratory depression.[4,5]

Halogenated, hypnotic, and opioid anesthetics and neuromuscular blockers may induce hypoventilation and respiratory failure. Non-depolarizing neuromuscular blockers have an unpredictable prolonged effect and the use of anticholinesterases requires caution. Even propofol, the most commonly used hypnotic for both anesthetic induction and maintenance, may present problems such as myotonia triggering and prolonged recovery.[1]

Due to the increased risk of malignant hyperthermia, depolarizing muscle relaxants, as well as halogenated ones, were avoided. Succinylcholine could also promote hyperpotassemia sufficient to lead to cardiac arrest or cause a generalized myotonic response, resulting in difficult orotracheal intubation and ventilation.

Phrenic nerve palsy is a well-known risk in both interscalene and cervical plexus block. This paralysis characterizes a potential risk for those with Steinert's disease who already have a respiratory deficit. Upper trunk block and selective supraclavicular nerve block were an option to interscalene and cervical plexus block for this patient, because the distance from the phrenic nerve in these sites is greater, which reduces the possibility of paralysis.[2,3] According to Kessler et al., at the cricoid cartilage level (C6), the phrenic nerve is 2 mm from the brachial plexus and at every 1 cm in the caudal direction, 3 mm are added to this distance.[6]

The beach chair surgical position for clavicle osteosynthesis impairs intraoperative access to the airway and brings a lot of discomfort to the sedated or awake patient, often leading to movements that interfere with the surgical procedure. In view of this surgical peculiarity in patients with clavicle fracture and Steinert's disease, it was considered more prudent to associate general venous anesthesia with the use of laryngeal mask, which allows a greater control of the airway. In the present case, the use of the laryngeal

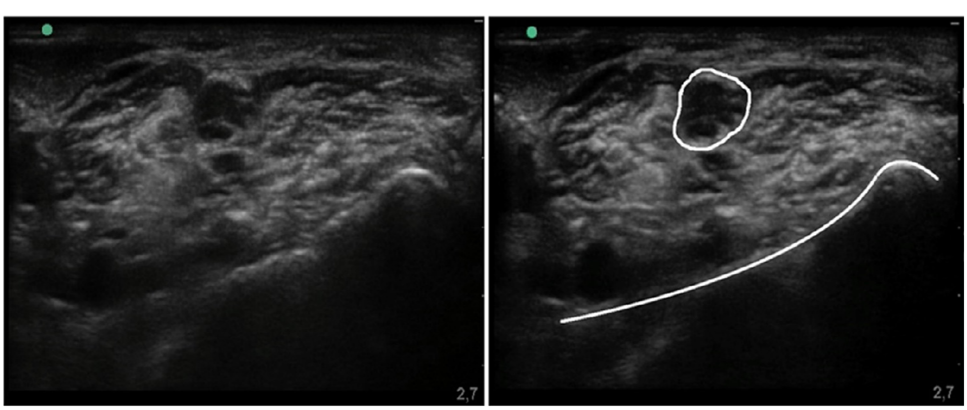

Figure 2 Merging of C5 and C6 for upper trunk formation in the center (circled to the right). Below, at a deeper level, the acoustic shadow of the first rib (contour in figure on the right) is already visible.

mask eliminated the need of using neuromuscular block and opioids for its introduction, allowing ventilation control and a calmer awakening.

Summarizing, sensory innervation of the clavicle remains controversial and still requires further studies to establish its precise origin. Above all, in this case report, the technique of supraclavicular nerve block associated with upper trunk blockade proved to be effective for the clavicle fracture osteosynthesis surgery, as maintenance of intraoperative hypnosis was the only requirement. In addition, the use of ultrasound in regional anesthesia allows increasingly selective blocks and promotes greater patient safety and lower morbidity. Considering the peculiarities imposed on the patient with Steinert's disease, the anesthesiologist must be aware of the preventive measures for myotonic crisis, peculiarities of the disease, and possible anesthetic techniques that aim to provide a safe condition during the preoperative period.

Conflicts of interest

The authors declare no conflicts of interest.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.bjane.2018.06.011.

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