

Anesthetic and surgical management of tracheotomy in a patient with Kennedy's Disease

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Abstract

Introduction. Kennedy's disease (KD) is a rare X-linked recessive motor-neuron neuropathy also known as Spinal and Bulbar Muscular Atrophy (SBMA).

Case report. We report a case of Kennedy's disease (KD) with acute laryngeal stridor and dysphagia whose anesthesiologic management required a tracheotomy.

The surgical procedure was conducted under local anesthesia performed by ultrasound-guided superior laryngeal nerve block and superficial cervical plexus block using 2% lidocaine. The surgical procedure was well tolerated without any discomfort reported from the patient.

Conclusions. In KD local anesthesia should be preferred to general anesthesia as general anesthesia is associated with increased potential for laryngospasm, pulmonary aspiration, and respiratory insufficiency related to the use of neuromuscular blocking agents and opioids.

At the best of our knowledge, this is the first case of tracheostomy in KD patient under locoregional anesthesia performed thorough superior laryngeal nerve and superficial cervical plexus block. *Clin Ter 2022; 173 (6):503-506 doi: 10.7417/CT.2022.2470*

Key words: Anesthetic Techniques, Kennedy's Disease, Cervical plexus block, Superior Laryngeal Nerve block, Tracheotomy

Introduction

Kennedy's disease (KD) is a rare X-linked recessive motor-neuron neuropathy also known as Spinal and Bulbar Muscular Atrophy (SBMA). It is caused by a Cytosine-Adenine-Guanine (CAG) repeat expansion of the first exon of androgen receptor (AR) gene. Several studies shows that the CAG repeats size correlates inversely with disease onset, but not with disease progression or severity. SBMA has an adult-onset, affect male and leads to limb and bulbar muscle atrophy, weakness and fasciculations (1). The most common signs and symptoms of bulbar involvement, present in 80% of cases, are tongue atrophy, vocal-cord paresis leading to dysarthria or dysphonia (2) and pharyngeal paresis

resulting in dysphagia (3). Spontaneous laryngospasm has been described, but its pathogenesis is still unclear (4-5). Respiratory failure is rare, although respiratory muscles may be involved at late stage of the disease (4). Non-neuronal symptoms, related to the partial loss of AR function, such as gynecomastia, can appear in the late stages of disease (6). Given the characteristics of KD, there are several potential risks related to the use of anesthesia such as laryngospasm, pulmonary aspiration (7) and also risks related to the use of neuromuscular blocking agents. Patients affected by KD have increased sensitivity to non-depolarizing neuromuscular blocking agents and succinylcholine (the only depolarizing neuromuscular blocking agent) may cause dangerous hyperkalemia (8).

Herein we report the case of a patient with KD that underwent a tracheotomy under locoregional anesthesia (LA) performed by ultrasound guided procedure and discuss the anesthetic technique and its potential clinical benefits. This case report has been prepared in accordance with CARE guideline. Written and informed consent for publication was obtained from the patient.

Case report

A 71-year-old man with acute laryngeal stridor was transferred to the Emergency Department of our University Hospital. The patient clinical record reported hypertension, chronic obstructive pulmonary disease (COPD), Kennedy Disease (KD) since 2004 and recent NSTEMI three months before needed coronary stents. The pharmacological therapy was Clopidogrel 75 mg once a day and Acetylsalicylic acid 100 mg once a day.

The physical examination shown stable vital signs: blood pressure 130/90 mmHg, 82 bpm heart rate and SpO₂ 97 % in the ambient air. All laboratory data were within normal limits: Hb 14.4 g/dl, PTLs 324.000 cell/mm³, glycemia 58 mg/dl, creatinine 0.5 mg/dl and troponin 0.104 mcg/l. The chest X-ray showed an accentuation of the broncho-vascular texture and bilateral hilar congestion.

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An otolaryngology examination with a fibro-laryngoscopy showed in the glottic site of the larynx a narrowing of the respiratory space and surgical tracheotomy was recommended.

Following the administration of 1 mg intravenous of midazolam, we positioned the patient in supine position and performed the following ultrasound (US) guided procedures: bilateral block of both superior laryngeal nerve (SLN) and superficial cervical plexus (SCP).

SLN procedure description

The SLN originates from the vagus nerve and splits in two branches: the external motor branch innervates the cricothyroid muscle of the vocal fold (9) and the internal sensory branch provides sensory to the mucosa from the base of the tongue down to the upper aspect of the vocal folds and functions as the afferent limb of the cough and laryngeal spasm reflexes (10). The superior laryngeal nerve space is identifying by placing the transducer in the sagittal plane to visualize the hyoid bone, the inferior border of great horn and the superior horn of the thyroid cartilage (Fig. 1). Subsequently the thyroid cartilage, thyrohyoid membrane and internal branch of superior laryngeal nerve space were identified. Therefore 4 ml of 2% lidocaine was injected and the SLN block was performed on both sides. SLN block was performed in preparation for a possible awake endotracheal intubation with the fibro-bronchoscopy.

SCP procedure description

After this first procedure, SCP block was performed. The SCP originates from the anterior rami of cervical spinal nerves C1-C4 and gives 4 terminal branches – lesser occipital nerve, transverse cervical nerve, supraclavicular nerve and great auricular nerve - for the sensory innervation of the skin and superficial structures of the anterolateral neck, external ear and shoulder (11). The SCP is located at

the posterior border of the SCM muscle at the level of the superior pole of the thyroid cartilage. The US transducer is placed in a transverse orientation across the neck and the needle is then inserted at the posterior border of the SCM at the level of the cricoid cartilage (12). Local anesthetic is injected deep to the SCM but superficial to the prevertebral fascia in order to avoid deep cervical plexus block (Fig. 2). SCP block was performed to numb the area around the first and second tracheal rings, where the tracheotomy was done. The patient lied in a supine position with head turned to the contra-lateral side for SCP block, then 6 ml of 2% lidocaine, was injected from both sides.

There were no complications during both blocks and the structures mentioned above were easily visualized due to the anatomy of the patient's neck (Fig. 3). Total time of the procedure was of ten minutes.

During the procedure, 100% oxygen was administered through a facemask at 4 liter/min and 50 mcg intravenous of fentanyl were administered in response to patient-reported discomfort. Patient's vital signs were stable during the surgical procedure: ECG was normal, blood pressure 144/80 mmHg, FC 79 heart rate and SpO2 100%. The arterial blood gas (A.B.G.) showed (FiO2 21%): pH 7,38, pCO2 44, pO2 96mmHg, lactate 0,6, HCO3- 28 mmHg.

At the end of the surgery the tracheostomy was correctly positioned, patient had no respiratory discomfort (Fig. 4). During the procedure, no LA by the surgeon was needed. The postoperative course was uneventful, and discomfort at the tracheostomy site was treated with 1 gr intravenous of acetaminophen.

Discussion

A Medline and Pubmed search revealed only two previous articles of anesthetic management in KD patients. The first described the successful management of an internal urethrotomy for urethral stenosis by epidural anesthesia (13); in the second case the authors reported the uneventful

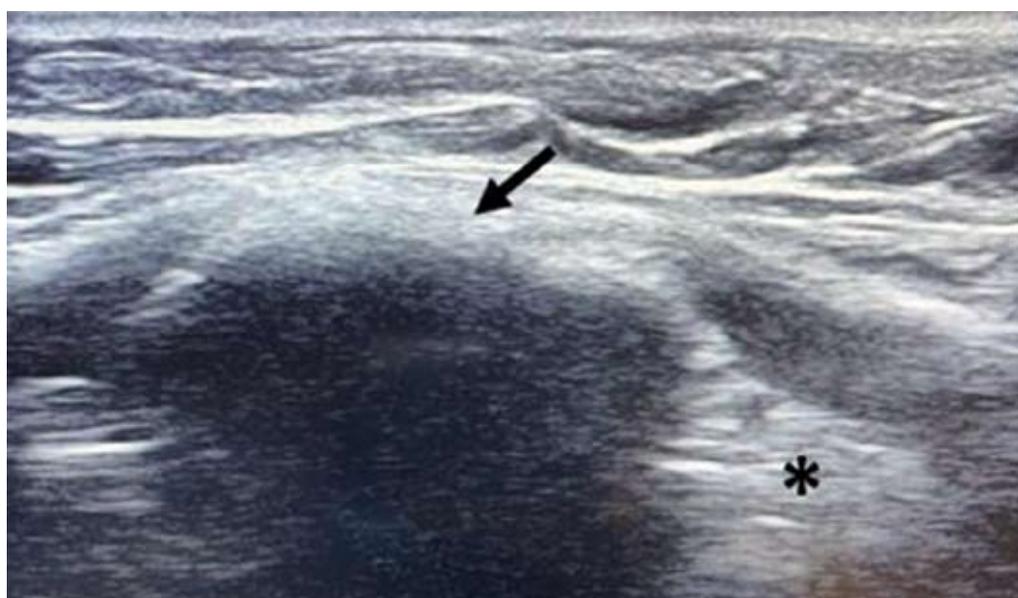


Fig. 1. The figure shows the hyoid bone (arrow) and the horns of the hyoid bone (asterisks), near which the superior laryngeal nerve is located.

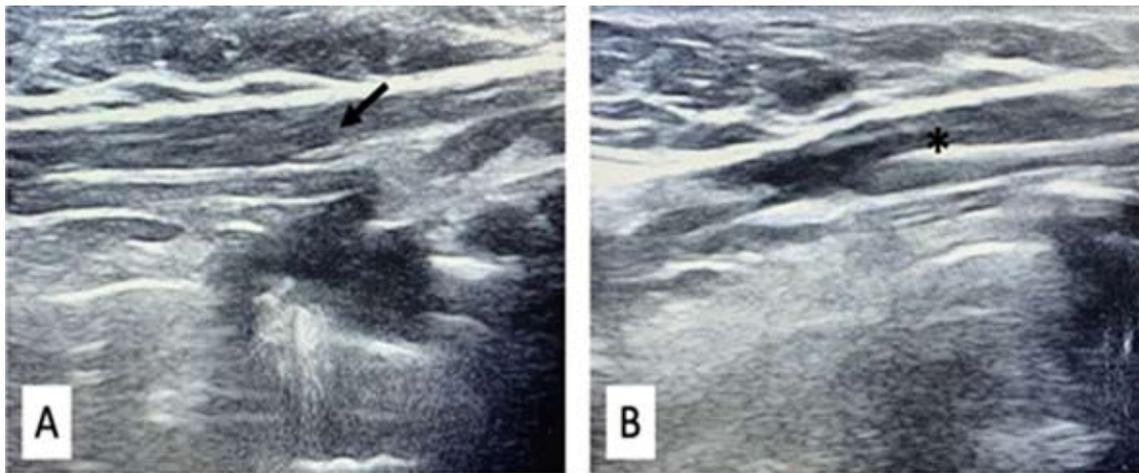


Fig. 2. A) Triangle of the sternocleidomastoid muscle (arrow). B) Needle for injection of the anesthetic deep to the muscle belly but superficial to the deep fascia (asterisks).

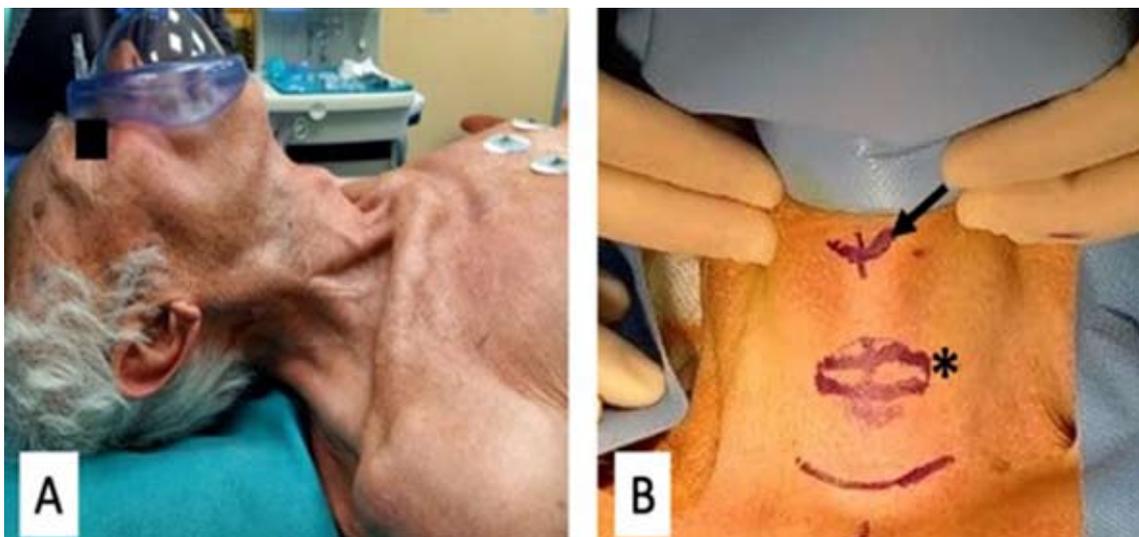


Fig. 3. Different orientation of the patient's neck. A) Lateral view B) Frontal view with thyroid (arrow) and cricoid (asterisks) cartilages of the larynx.



Fig. 4. The end of the surgery and position of the tracheostomy tube.

administration of general anesthesia without neuromuscular blocking agents and no incidents of laryngospasm (14).

This is the first described case of tracheotomy performed in locoregional anesthesia in a patient with KD. We chose LA ultrasound guided due to block SLN and SCP with 2% lidocaine for several reasons. First of all, SLN and SCP block have been described to effectively anesthetize both the larynx and the patient's neck for airway manipulation, surgical treatment on the neck (15) and pain control (16). Furthermore, we adopted LA to avoid interactions between anesthetic drugs like neuromuscular blocking agents and opioids. In fact in this type of patients, the management with neuromuscular blocking agents should be avoided because was observed an increase sensitivity to non-depolarizing neuromuscular blocking agents and succinylcholine is contra-indicated in this patient population due to the risk of lethal hyperkalemia (8). Moreover, regional approaches may lower the risk of post-operative respiratory depression (17).

The ultrasound guided SCP block has multiple advantages: it is easy to perform and allows the spread of LA in the correct plane of the neck. The major complication of the SCP block is injection of local anesthetic under prevertebral fascia leading to blockade of the deeper neural structures like the cervical plexus, brachial plexus and the recurrent laryngeal nerve. The most important complication in patients with neuromuscular disease after SCP is the blockade of phrenic nerve, especially in bilateral procedure, due to its effect on diaphragm functionality in those patients at high risk of respiratory impairment.

Conclusions

KD is a rare syndrome and few articles in the scientific literature are available. In this case, we successfully managed a patient with KD by SLN and SCP blocks before surgical tracheotomy. Clear guidelines for anesthetic management in KD patients are not available, then we suggest that in KD patients, where is possible, a regional anesthetic is required, to avoid manipulation of the airway.

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