OPIOID-SPARING ANESTHESIA AND CHARCOT-MARIE-TOOTH DISEASE: A CASE REPORT

Mongiovì N[12], Vignali A[1], Bertoni G[12], Guadagnucci M[12], Baratta A[1]

[1]Anestesia Rianimazione ed Elisoccorso - Ospedale Apuane - Azienda USL Toscana nord ovest

[2]Azienda Ospedaliera Universitaria Pisana

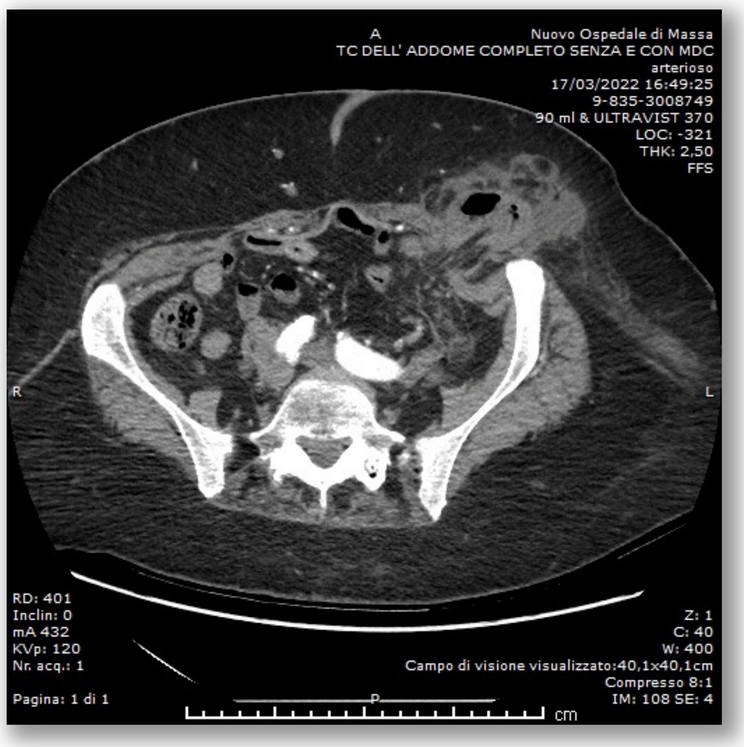
Background:

Charcot-Marie-Toot (CMT) disease is the most common inherited motor-sensory disorder characterized by:

- ✓ Mutations in genes encoding neuronal proteins, most of which affecting the **myelin sheath**, in other cases axonal transport or mitochondrial metabolism.
- ✓ Muscle **wasting** and weakness start in the lower limbs and slowly spread cranially involving hands and forearms, severe cases may be characterised by muscular **atrophy** of the respiratory muscles, diaphragm and vocal cords. **Autonomic dysfunction** is also commonly present.
- ✓ Anaesthetic literature is not clear about the best anaesthetic options for patients with CMT: there are many **concerns** about delayed onset of action of muscle relaxant drugs, hyperkalaemia, malignant hyperthermia, difficulties with positioning, risk of nerve injury using regional anaesthesia techniques and postoperative respiratory insufficiency.

Case Report:

58-year-old woman diagnosed of Charcot-Marie-Tooth disease underwent **laparoscopic incisional hernia repair** in general anaesthesia. As for her diagnosis she complained little weakness in the lower limbs without strength deficit in the upper limbs. She suffered also from obesity, arterial hypertension, previous episodes of retinal thrombosis.
Our main **concerns** were the optimal neuromuscular block management and the risk of respiratory insufficiency after surgery.



Anaesthesia induction and maintenance:

Anaesthesia induction → Propofol 150 mg, Fentanyl 50 mcg and Rocuronium 40 mg.
General anaesthesia maintenance→ Sevoflurane MAC 1 and target-controlled-infusion of Remifentanil (1.5 ng/ml Cpt).
Neuromuscular block maintenance→ We monitored Train-of-Four and Post-Tetanic-Count on the right ulnar nerve. 10 mg of Rocuronium 80 minutes from the induction were the only additional dose required to maintain optimal laparoscopic conditions.

At the end of surgery:

- ✓ Paracetamol 1 g plus Ketorolac 30 mg iv.
- ✓ Ultrasound-guided bilateral Transversus Abdominis Plane (TAP) Block using a lateral approach and infusing 40 ml (20 ml per side) of a solution containing Ropivacaine 100 mg and Mepivacaine 200 mg.
- ✓ Sugammadex 200 mg to reverse neuromuscular block.

During the one-hour after-surgery monitoring and hospitalization no opioid-pain control therapy was required. She was discharge on post operative day 2.

Our Experience:

Use of Sevoflurane

The concern that CMT had a higher risk of **malignant hyperthermia** is based upon previous categorisation as a muscular dystrophy: now we know that this is a peripheral neuropathy, not a myopathy, and this risk appear to be unfounded [1-2].

→ We took advantages of its **potentiating effect** on muscle relaxation in order to reduce Rocuronium dose.

Use of Rocuronium

Variable response expected depending on motor unit loss and upregulation of acetylcholine receptors in the neuromuscular junction. Some CMT patients have increased **sensitivity** to non-depolarizing neuromuscular blockers, in other cases other may show normal or moderate **resistance**.

→ In our experience Rocuronium efficacy was **prolonged**, we needed one additional bolus after 80 minutes from the induction to maintain a PTC < 3. **Sugammadex** response was efficient and quick, obtaining a TOF Ratio > 90% in few minutes after the administration.

Use of Opioid

CMT patients can be at risk of pulmonary complications after surgery [3] such as **respiratory insufficiency** or pneumonia for multiple causes (thoracic muscles and diaphragm weakness, restricted respiratory dysfunction).

→ We decided for an **opioid free strategy** to minimize respiratory complications' risk without any trouble in achieving optimal pain control.

TAP Block

There are concerns using regional techniques in CMTD, patients may be more susceptible to **nerve injury** [4]. Nerve blocks and neuraxial blocks have been performed in patients with stable CMTD for anesthesia and analgesia without adverse neurologic outcomes. CMTD patients may also have an increased susceptibility to local anesthetic neurotoxicity, which is both concentration and dose dependent.

→ **Reduction** in local anesthetic concentration and dose is advisable and we used **ultrasound-guided** technique to facilitate the precision of local anesthetic injection, reducing drug doses. In our experience it was well-tolerated and no adverse effect occurred.

We hypothesized that an underlying low degree of **chronic sensory and pain impairment** may contributed to the optimal postoperative pain control.

1. Antognini JF. Anaesthesia for Charcot-Marie-Tooth disease: a review of 86 cases. Can J Anaesth. 1992 Apr;39(4):398-400
2. Pasha TM, Knowles A. Anaesthetic management of a patient with Charcot-Marie-Tooth disease for staged diaphragmatic plication. Br J Anaesth. 2013 Jun;110(6):1061-3
3. Aboussouan LS, Lewis RA, Shy ME. Disorders of pulmonary function, sleep, and the upper airway in Charcot-Marie-Tooth disease. Lung. 2007 Jan-Feb;185(1):1-7
4. McSwain JR, Doty JW, Wilson SH. Regional anesthesia in patients with pre-existing neurologic disease. Curr Opin Anaesthesiol. 2014 Oct;27(5):538-43