

SPINAL ANAESTHESIA IN A PATIENT WITH MACHADO-JOSEPH DISEASE

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Introduction

Machado Joseph Disease (MJD) or spinocerebellar ataxia type 3 (SCA3) is an inherited neurodegenerative disorder characterized by progressive sensory and motor dysfunction. MJD is a rare disease and to the extent of our knowledge only 2 reports document the anaesthetic management of patients affected by this condition: one used an epidural anaesthesia for inguinal hernia repair and the other used a combined spinal-epidural technique with hyperbaric bupivacaine for vaginal hysterectomy. We describe the third case.

Clinical Case

A 65-year-old woman with MJD complicated with severe weakness in the arms and legs and diffuse polymyalgias presented in the emergency setting for surgical repair of a bimalleolar fracture. Besides the neurodegenerative disorder she also had atrial fibrillation, bradycardia-tachycardia syndrome with an artificial pacemaker in place, hypertensive disease, obesity and dyslipidemia. After informed consent, a subarachnoid block was performed at the L3-L4 lumbar intervertebral space with 10 mg of hyperbaric bupivacaine taking advantage of an expected lateralization of the blockade due to the positioning and so a more expectable cardiovascular stability. Sensory block reached the T10 level. The procedure was uneventful. Postoperative normal recovery occurred and the patient was discharged 2 days after. One month after the spinal anaesthesia no new neurological changes have been identified or reported when compared to the preoperative setting.

Drugs	
Fentanyl IV	0.10 mg
5mg/ml hyperbaric Bupivacaine	10.00 mg
Cefazoline IV	2000.00 mg

Discussion

Central nervous system disorders may be considered a relative contraindication to regional anaesthesia because of the risk and difficulty in determining the cause of eventual exacerbation of neurological deficits. Nevertheless, given that there are no large clinical studies supporting either type of anaesthetic management in MJD, we opted for regional blockade instead of general anaesthesia, taking into consideration the 2 uneventful cases already reported in literature and the cardiac disease of the patient. The regional technique allowed hemodynamic stability, an easier postoperative analgesic management and minimized post-procedure pulmonary complications such as hypoventilation, aspiration and hypoxia due to impaired neuromuscular function.

Learning points...

Neuroaxial anaesthesia with hyperbaric bupivacaine can be a safe option for lower limb surgery in patients with MJD.

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