453545 - SPINAL ANAESTHESIA IN A PATIENT WITH KENNEDY'S DISEASE

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Purpose: In this report, the authors present the anaesthetic management of a 43 year old man with Kennedy's disease who was scheduled to undergo orthopaedic surgery under spinal anaesthesia.

Clinical Features: Local REB approval was obtained for this study. A 43-yr-old man with Kennedy's disease was scheduled for open reduction and internal fixation of a left ankle lateral malleolar fracture. Four years ago, he developed a postural hand tremor, and a slight swallowing difficulty. He had been admitted to a local hospital for a work up on 2003. The physical examination revealed fasciculation and atrophy of the tongue and facial muscles, electromyography (EMG) showed large polyphasic motor units in affected muscles, and a laboratory examination showed that serum creatine kinase was elevated at 290 u litre⁻¹ (normal range 26-200 u litre⁻¹). DNA analysis confirmed Kennedy's disease by demonstrating a triplet (CAG) repeat expansion in the androgen receptor gene on X chromosome. Our preoperative assessment revealed that he was 165 cm tall and weighed 75 kg. His physical examination findings were normal, with the exception of mild swallowing problems, weakness of the lower and upper extremities, and fasciculations. Spinal anaesthesia was chosen after an extensive discussion of the risks and benefits involved. On arrival in the operating room, non-invasive blood pressure was 130/85 mm Hg, heart rate 80 beats min⁻¹, and oxygen saturation 98%. Following the rapid administration of 500 ml of Ringer's solution intravenously, spinal anaesthesia was performed at the L4-L5 interspace using a 25-G Quincke type needle. Bupivacaine hydrochloride (2 ml; Marcaine® 0.5% spinal, AstraZeneca) was administered. Surgery was started after achieving anaesthesia at the T10 dermatome. Postoperatively, his neurologic status remained normal with no worsening of his dysphagia or extremity weaknesses. He was discharged from hospital without any neurologic complaint on the sixth postoperative day.

Conclusion: Kennedy's disease typically presents as muscular atrophy, weakness, and fasciculations predominantly of bulbar, facial, and proximal muscles of the extremities. When a patient with Kennedy's disease is scheduled to undergo a procedure requiring anaesthesia, anaesthesiologists should carefully assess patient preoperative status, respiratory function, and inquire about swallowing difficulties or a history of intolerance to any general or local anaesthetic agent. Considerations for general anaesthesia concern possible prolonged neuromuscular blockade and consequent postoperative muscle weakness and a compromised baseline pulmonary function.¹ Many inhalation agents have been reported to be well tolerated by the patients with lower motor neuron disease.² However, it is not clear whether neuromuscular blocking agents prolong neuromuscular blockage. Depolarizing muscle relaxants, such as, succinylcholine, are not recommended. Moreover the possibility of hyperkalemia and of resultant ventricular arrhythmia or fibrillation has been reported in patients with neuromuscular and lower motor neuron

diseases by several investigators.³ If muscle relaxation is required, non-depolarizing neuromuscular blocking agents should be considered, and neuromuscular transmission should be monitored closely.²

References: 1. Int J Obstet Anesth 2004; 13: 192-5 2. Clinical Neuroanesthesia. New York: Churchill Livingstone, 1990; 351-77 3. JAMA 1970; 213: 1867-71