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Research Paper

Anaesthetic Considerations in A Patient With Limb Girdle Muscular Dystrophy For Fracture Supracondylar Femur

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I. INTRODUCTION

Limb-girdle muscular dystrophy (LGMD) refers to a genetically heterogeneous group of muscular dystrophies that present with weakness mainly involving the shoulder and hip girdles. LGMD has a predominantly proximal distribution of weakness. In the early course of the disease the distal, facial and extra-ocular muscles are spared[1]. The adult-onset disease involves both shoulder and pelvic girdles with gradually increasing proximal limb weakness. This leads to restriction of mobility and eventually leads to wheelchair confinement. The muscles most affected are those closest to the body (proximal muscles), specifically the muscles of shoulders, upper arms, pelvic area, and thighs[2]. It is difficult to determine the prevalence of limb-girdle muscular dystrophy because its features vary and overlap with those of other muscle disorders. Some factors such as severity, age at onset, and features of LGMD are varied among patients. Anaesthetic complications/implications in these patients are secondary to effects of anaesthetic drugs on myocardial and skeletal muscles. Events such as cardiac arrest, malignant hyperthermia (MH) and delayed recovery from non-depolarising muscle relaxants pose a challenge to the anaesthesiologist[3].

II. CASE REPORT

A 45 year old male weighing 69 kg with myotonic dystrophy was scheduled for open reduction and internal fixation (Rt) for bilateral supracondylar femur fracture. He was diagnosed with LGMD at age of 30 years with electro-myography showing signs of early myopathy and muscle biopsy showing subtle myopathic changes and myeloid degeneration. Creatinine phosphokinase (CPK) level was raised (396 U/L). His symptoms predominantly affected proximal muscles due to which, initially he had difficulty in getting up from a sitting posture and had a waddling gait. He was on wheel chair for the last 6 years.He had no cranio-bulbar symptoms. History of similar illness was present in the family (sibling bedridden from similar illness). He had prior anaesthetic exposure was for surgery 4 year back and perioperative status was uneventful. On examination patient was conscious and oriented with stable vitals. Cardiovascular system examination noticed ejection systolic murmur in aortic area. Motor system examination revealed upper limb power of 4/5 and lower limb power of 1/5.Sensory system was within normal limits. Difficult airway was ruled out. Other pre-operative investigations were within normal range. Echocardiography showed evidence of non-obstructive hypertrophic cardiomyopathy. Operating room was prepared according to malignant hyperthermia protocol. Aspiration prophylaxis was given. Standard monitors were attached and baseline vitals recorded (pulse-100 bpm, blood pressure-140/80 mm of Hg, respiratory rate-18/min, temperature-37.1°C and SpO_2 -100% at room air). Eighteen-gauge intravenous line was secured, Ringer lactate infusion started and injection midazolam 1 mg intravenously was administered. Combined spinal epidural block was given with 1.0ml heavy bupivacaine 0.5% with 25 mcg fentanyl in sitting position. The procedure was started after a block level of T10 open reduction and internal fixation was completed uneventfully with stable intra-operative haemodynamic and SpO2 of 99–100% on oxygen face mask. After the surgery, the patient was shifted to recovery room. The block receded completely over the next 4 h.

III. DISCUSSION

Anaesthesia in patients with neuromuscular diseases is a great concern for anaesthesiologists. LGMD are a heterogeneous group of genetically determined progressive disorders of skeletal muscles with both autosomal dominant and recessive inheritance. They are characterised by proximal muscular dystrophy, elevated creatine kinase and associated cardiorespiratory problems.

The anaesthetic considerations of LGMD are similar to other muscular dystrophies. However, perioperative complications are not proportional to the severity of the disease and occur even in mildly affected patients which need careful pre-operative evaluation and consultation.

Regional anaesthesia should be performed whenever possible as general anaesthesia in LGMD needs careful monitoring, due to the high incidence of fatal complications. In LGMD, cardiac muscle and conducting pathways are affected and the sudden appearance of a Wenckebach type of block can occur especially during change in patient's position. In the absence of cardiomyopathy, propofol and thiopentone can be safely used as induction agents. Respiratory compromise may occur early due to severe diaphragmatic involvement resulting in hypoventilation. Hence, sedative-hypnotics and opioids should be used judiciously. In patients with muscular dystrophies, suxamethonium and to a lesser extent volatile anaesthetics should be avoided as they may lead to life-threatening complications such as rhabdomyolysis and malignant hyperthermia. These patients are sensitive to non-depolarising agents and it is recommended to use titrated doses of rocuronium and atracurium under neuromuscular monitoring. Inhalational, depolarising and non-depolarising muscle relaxants (being triggering agents in MH and also with risk of respiratory insufficiency) needed to be avoided[4].

General anaesthesia has to be considered in patients who are unable to tolerate supine position despite respiratory support, or patients having bulbar muscle involvement. Our patient had weakness only in proximal muscles and no cardio-respiratory involvement. Hence, we planned to give spinal anaesthesia to our patient with arrangements for general anaesthesia considered if required. Additionally, we avoided general anaesthesia because the use of anaesthetic agents in these patients make them more susceptible for malignant hyperthermia and excessive sedation with possibility of post operative respiratory complications .Regional nerve block was another option for this patient but difficulty in positing this patient make this method difficult(patient had bilateral fracture). In addition to effect of anaesthetic agents, spinal anaesthesia with high volume of drug in patient with hypertropic cardiomyopathy added additional risk of hypotension further reducing afterload and cardiac output causing cardiovascular compromise[5].

Limb girdle muscular dystrophy patients are confined to bed and are more prone for the development of deep vein thrombosis which can be prevented by use of compression stockings or sequential compression devices intraoperatively and low molecular weight heparin prophylaxis.

IV. CONCLUSION

Management of limb girdle muscular dystrophy should be individualized as the symptoms vary among patients. Severe cases are best managed by multi-disciplinary team. In this case report we highlight the fact that combined spinal epidural anaesthesia can be safely used to provide anaesthesia for lower limb surgeries in patients with limb girdle muscular dystrophy. Further studies are required to establish definite anaesthetic management strategies as few references are available for this rare disease.

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