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Case Report

Anaesthetic management of a patient with Becker's muscular dystrophy undergoing orthopaedic surgery: A case report

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Abstract

Becker muscular dystrophy (BMD) is an inherited congenital muscular disorder caused due to mutation in the dystrophin gene. Clinical presentation includes progressive muscle loss and variable weakness involving primarily the proximal lower limb muscles. BMD has a milder disease course compared to Duchenne Muscular dystrophy; but as the disease progresses patients invariably develop cardiac failure and less commonly respiratory failure. Anaesthetic management of patients with BMD are challenging to the anaesthesiologist due to the risk of malignant hyperthermia, rhabdomyolysis, hyperkalaemia, cardiac arrhythmia and respiratory failure.

Keywords: Becker muscular dystrophy, surgery, orthopaedic, anaesthesia, spinal, ropivacaine

Introduction

Becker muscular dystrophy (BMD) is a rare, inherited degenerative muscular disorder that is almost exclusively seen in males. BMD arises from a mutation in the sub sarcolemmal protein dystrophin which is located in the Xp21.2 chromosome [1].

The prevalence of BMD is 17-27 cases per 1 million population and the estimated incidence of BMD is 1 per 30,000 male births in the United States [2]. In Asia the prevalence of BMD is 0.1 to 0.2 per 10,000 male individuals [1]. Compared to Duchenne muscular dystrophy (DMD), BMD is characterized by a delayed and milder presentation. The clinical manifestations of BMD occur widely between 5 and 60 years of age [1]. The clinical features includes progressive muscular weakness and wasting, involving initially the proximal lower limb muscles and pelvic muscles, and then progressing to involve the muscles of the arms, shoulder, and neck. Pseudohypertrophy of the calf muscles occurs frequently. Approximately 90% of patients with BMD develop disorders of both conduction and contractility of the heart, leading to dilated cardiomyopathy, which remains the primary cardiac involvement [3].

In patients with a family history of BMD, the diagnosis is straight-forward, but in cases of sporadic illness, genetic analysis should be performed [1]. Diagnosis of BMD is now done using DNA testing of the dystrophin gene, whereas earlier the diagnosis was based on muscle biopsy with dystrophin staining [4]. However, although glucocorticoid therapy has shown to slow disease progression, there is no effective treatment currently [1].

Patients with BMD who are exposed to anaesthesia are at risk of developing perioperative complications such as hyperkalaemia, rhabdomyolysis, malignant hyperthermia, congestive cardiac failure, dysarrythymia, and respiratory depression. Exposure to succinylcholine and halogenated agents during anaesthesia can lead to fatal atypical reactions in anaesthesia and even sudden cardiac arrest [5]. Hence, patients with BMD need a thorough preoperative evaluation, meticulous intraoperative anaesthetic management, and good postoperative care. We report a case of a 19-year-old male who is a known case of Becker muscular dystrophy and presented with a fracture shaft of the left femur, scheduled for closed reduction and internal fixation of the left femur fracture.

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Case Report

A 19-year-old male patient was admitted to the orthopaedic department with an alleged history of slip and fall, following which he sustained injury to the left lower limb. On radiological examination, the patient was diagnosed to have a fracture of the shaft of left femur and was scheduled for closed reduction and internal fixation with intramedullary nailing the following day. On history taking, the patient was diagnosed to have Becker muscular dystrophy at the age of 13 years, presenting initially with muscle weakness that affected his ability to climb stairs. The diagnosis was confirmed by a muscle biopsy. There was also a family history of BMD. There was no previous anaesthesia exposure or history of anaesthetic complication. On examination, the patient had pseudo hypertrophy of both the calf muscles and bilateral pes cavus deformity. The patient underwent a thorough pre-anaesthetic evaluation to know the extent of the disease and cardiovascular and respiratory system involvement. All the routine investigations were within normal limits, including chest x-ray, electrocardiogram and 2D-ECHO.

Prior to surgery, the patient was kept nil per oral for 6 hours. On the day of surgery, in the preoperative room, standard American Society of Anaesthesiologist (ASA) monitors were attached and an 18-gauze intravenous (IV) cannula was secured in the dorsum of left hand. Preloading was done with 250ml of ringer lactate solution over 20 minutes. In the operating room, all the airway gadgets and emergency drugs were kept ready. Vaporizers were removed from the workstation, and the breathing circuits were flushed with oxygen multiple times to avoid the risk of malignant hyperthermia. A subarachnoid block was planned for the proposed procedure in our patient. After attaching the standard ASA monitors and under strict aseptic precautions, in sitting position, a lumbar puncture was performed at L3-L4 level using 27 Gauze Quincke's spinal needle. After confirming the clear and free flow of cerebrospinal fluid, Inj. Ropivacaine Heavy 0.75% 3 ml with adjuvant buprenorphine 60 mcg was injected intrathecally. After achieving adequate sensory and motor blockade surgery was started. The surgical procedure completed within the duration of 2 hours. Throughout surgery, the patient was hemodynamically stable, and surgery was uneventful. The patient was then shifted to the postoperative room and monitored for 4hours. Post-operative analgesia was maintained with the help of injection Tramadol 50mg IV BD and injection Paracetamol 1gm IV TID. After complete recovery from motor (according to modified bromage scale) and sensory blockade patient was shifted to the ward. Recovery was uneventful, and on the 5th postoperative day, the patient was discharged.

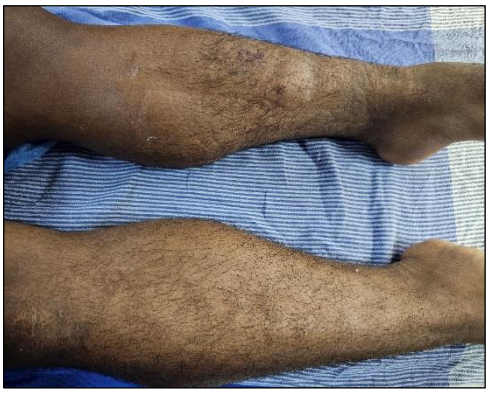

Fig 1: Bilateral psueudo-muscular dystrophy of calf muscles

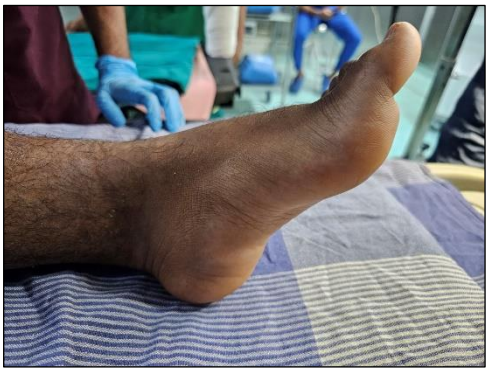

Fig 2: Cavus deformity of foot

Discussion

Becker muscular dystrophy is an X-linked congenital muscular disorder characterized by the absence of the dystrophin gene due to the deletion mutation in the Xp21.2 chromosome [1]. Compared to Duchenne muscular dystrophy, BMD has a late presentation (average age of presentation being 12 years) and has a

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benign course. As the disease progresses, patients invariably develop respiratory and cardiac failure since there is no cure. Through medical intervention, if the cardiac symptoms are adequately controlled, a near-normal life span can be expected in these patients [6].

Patients with BMD who are posted for surgery pose a challenge to the anaesthesiologists as it is associated with complications that can be life-threatening. The anaesthetic complications of BMD include hyperkalemia, malignant hyperthermia, rhabdomyolysis, dysarrthymia, cardiac failure, and respiratory failure; hence, patients need a thorough pre-anaesthetic evaluation and preparedness before any surgical procedure. Irrespective of the age, signs and symptoms, most patients with neuromuscular disorders have associated cardiac or respiratory system involvement and must undergo cardiac evaluation to rule out occult cardiomyopathies [7]. Compared to patients with DMD, those with BMD are at high risk of developing dilated cardiomyopathy leading to cardiac failure. Whereas, some patients may have fragile muscles that break down under the stress of surgery or anaesthesia. The stress of surgery, pressure due to positioning for surgery, application of a tourniquet, and postoperative shivering may all contribute to complications in patients with muscular dystrophies [7]. They may also develop unexpected adverse reactions to anaesthetic drugs [6].

In patients scheduled for surgery, there is no definite recommendation for anaesthesia [8]. Both regional and general anaesthesia are challenging in patients with BMD.

In certain types of surgeries, general anaesthesia (GA) is unavoidable. GA is associated with various complications such as aspiration pneumonia, delayed recovery, prolonged mechanical ventilation, and postoperative shivering. Patients with BMD are extremely sensitive to depolarizing muscle relaxant (succinylcholine) and can produce prolonged contraction even though the risk of malignant hyperthermia is thought to be less as compared to DMD. Upon administration of succinylcholine, the patient might develop hyperkalaemia secondary to rhabdomyolysis, resulting in cardiac arrest. So, whenever in need, a nondepolarizing muscle relaxant can be administered along with neuromuscular monitoring. Use of volatile agents in these patients is associated with a risk of developing malignant hyperthermia. Therefore, inhalational anaesthetics should be avoided, and total intravenous anaesthesia can be recommended in these patients. With great caution, sevoflurane and desflurane can be administered by an experienced anaesthesiologist.

Central neuraxial blocks are considered a better option for patients with BMD [9]. Subarachnoid block (SAB) is associated with a lack of control over hemodynamic parameters, making meticulous monitoring mandatory. With the introduction of local anaesthetics with a better pharmacological profile, regional anaesthesia like SAB is now considered safe in patients with BMD. Ropivacaine is one such drug that is nearly identical to bupivacaine in onset, quality, and duration of sensory blockade, but it produces less motor blockade with less haemodynamic change and has a better safety profile.

In patients with muscular dystrophies, peripheral nerve blocks are also considered safe as they have minimal impact on the haemodynamics and respiratory function, thereby proving beneficial in patients with cardiorespiratory involvement. Moreover, peripheral nerve bocks provide effective postoperative analgesia without the need for additional drug usage [9].

Our patient presented with a fractured shaft of the left femur without significant muscular weakness; hence, regional anaesthesia i.e. SAB was preferred, considering the advantages over general anaesthesia such as nil airway manipulation, no risk of malignant hyperthermia, no risk of postoperative respiratory depression/postoperative nausea and vomiting, avoidance of polypharmacy, and a better safety profile of ropivacaine, along with good postoperative analgesia. But before administration of the SAB, the patient has to undergo neurological examination, cardiac evaluation, and vigilant administration of fluid in the perioperative period to avoid cardiac catastrophe. Irrespective of the type of anaesthesia we choose to administer, emergency drugs (vasopressors, ionotropes, antiarrhythmic agents, diuretics, etc.) must be kept ready in order to manage any perioperative complications.

Conclusion

In patients with Becker muscular dystrophy undergoing lower-limb surgeries under subarachnoid blocks, ropivacaine can be safely administered after a thorough pre-anaesthetic check-up with cardiac evaluation and meticulous intraoperative fluid management.

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