

Case report

Anesthetic Management of a Case of Duchenne Muscular Dystrophy in Congestive Heart Failure with Compartment Syndrome of the Upper Limb for Emergency Fasciotomy

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Abstract

We describe the effectiveness and safety of regional anesthesia in a patient with Duchenne muscular dystrophy (DMD) with a deranged coagulation profile. A 16-year-old male having DMD and congestive heart failure (CHF) presented to emergency with abdominal pain. Following cannula insertion, the patient developed compartment syndrome of the left forearm and hand. He was started on thrombolytic heparin infusion, and the surgical team planned for an emergency fasciotomy. Ultrasound-guided axillary plexus block with ropivacaine allowed us to provide adequate anesthesia for this patient avoiding the deleterious effects of general Anesthesia.

Keywords: Duchenne Muscular Dystrophy; Regional Anesthesia; Coagulation

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Introduction

Muscular dystrophy is a hereditary disorder characterized by atrophy and painless degeneration of skeletal muscles. Duchenne (DMD) is an X-linked recessive type of muscular dystrophy with 1 in 5000 boys (1). The complete or partial loss of dystrophin protein is the primary etiology, leading to disruption of sarcolemmal integrity, causing myofibril atrophy, necrosis, and fibrosis. The patients of DMD have proximal myopathy with Gower sign positive and progressive respiratory and cardiovascular systems involvement (1, 2). We describe the successful anesthetic management of a patient of DMD with congestive heart failure (CHF) on heparin infusion

with compartment syndrome posted for fasciotomy using an ultrasound-guided axillary brachial plexus block. Ropivacaine was the preferred local anesthetic for performing the block because it is less cardiotoxic (a pure S-enantiomer binds less firmly to the sodium channels) than racemic bupivacaine. Ropivacaine also has a higher cardiovascular collapse to central nervous system (CC/CNS) toxicity ratio. Therefore it has a broader safety margin as regards local anesthetic systemic toxicity (LAST) (3). In addition, it also has intrinsic dose-dependant vasoconstrictive properties that prolong the duration of action at the site of injection and prevent rapid systemic absorption of the drug, further decreasing chances of LAST (3).

Case Report

A 16-year-old, ASA physical status III, the male patient presented to the emergency department with abdominal pain. He was a previously diagnosed case of Duchenne muscular dystrophy (confirmed on genetic testing) with bilateral lower limb weakness and cardiomyopathy on follow-up with the Neurology department of our hospital. He was found to have raised jugular venous pressure, pedal edema, and hepatomegaly on present admission. Echocardiography revealed global hypokinesia with a left ventricular ejection fraction of 15-20%, mild mitral and tricuspid regurgitation, and right ventricular dysfunction leading to CHF. The patient was started on dobutamine infusion and tablet torsemide. In the next 24 hours, the patient developed pain and swelling in the left forearm and hand, with a feeble pulse compared to the right side. A Doppler study showed a thrombus in the left axillary, brachial, and radial vein; normal arterial flow. The patient was started on heparin infusion 1200U/hour, and as compartment syndrome could not be ruled out, the patient was scheduled for a fasciotomy.

Hematological investigations just before surgery revealed INR- 2.8, high activated partial thromboplastin time (APTT), and platelet count of 81,000/mm³; the remaining investigations were within normal limits. Since it was an emergency surgery, heparin was stopped patient was shifted to the operating room with 4 units of fresh frozen plasma. As the patient had cardiomyopathy with CHF and needed surgery on the forearm and hand, we decided to perform an ultrasound-guided axillary brachial plexus block for providing anesthesia. The procedure was explained to the patient, and his consent was obtained. After attaching standard monitors (5-lead ECG, SpO₂, skin temperature probe), an experienced anesthetist performed the axillary block using a short bevel 5cm block needle and 15ml of 0.5% ropivacaine. The surgery was started after confirming sensory anesthesia of the forearm and hand. The subsequent intraoperative course was uneventful, with blood loss of 150-200. The patient was then transferred to a high dependency unit for monitoring. He was discharged from the hospital 10 days later after features of CHF subsided. Consent was achieved before reporting the case.

Discussion

In patients with DMD, loss of dystrophin, a large protein that anchors the actin-myosin complex to the cell membrane and surrounding cellular matrix, results in ineffective muscle contraction after transmission of electric impulses. This results in progressive skeletal proximal myopathy and up-regulation and the development of extra-junctional acetylcholine receptors (1). On the cellular level, the instability caused by the loss of dystrophin results in increased permeability of the sarcolemma and increased intracellular calcium. Exposure of these patients to inhalational anesthetic agents or depolarising muscle relaxants increases intracellular calcium, leading to rhabdomyolysis, hyperkalemia, and cardiac arrest (2). Due to increased junctional neuromuscular receptors, these patients can also have exaggerated hyperkalemia in response to the administration of a depolarising muscle relaxant leading to fatal cardiac arrest (2). These patients also have an unpredictable response to non-depolarising muscle relaxants and need postoperative mechanical ventilation (2). The changes in dystrophin also extend to the cardiac and smooth muscles, leading to progressively worsening cardiomyopathy (1, 2). In addition, weakness of the diaphragm, intercostal and accessory muscles of respiration lead to restrictive pulmonary dysfunction, which increases the risk of respiratory failure and postoperative ventilator requirement (1, 2). These patients' primary cause of morbidity is cardiomyopathy and a higher risk of major adverse cardiac events despite normal pre-operative echocardiography reported with inhalational agents and succinylcholine (2). Though propofol-based total intravenous anesthesia has been safely used for patients of DMD, it has adverse inotropic effects, and the present patient presented with features of heart failure and needed inotropes and diuretics for the treatment of the same. In this situation, an ultrasound-guided axillary block was a safer option than general anesthesia (GA). The only concerns were that the patient was receiving thrombolytic medications. Though ultrasound may not prevent intravascular injection, the ASRA guidelines and a recent meta-analysis have elucidated that judgment to perform a peripheral nerve block should be based on how superficial the nerve block is, the

compressibility, vascularity, and consequences of bleeding (4, 5). Though deep blocks such as the lumbar plexus block should be avoided in patients on thrombolytic medication, an axillary block is superficial. It was at a depth of 1.5cm in this patient, and therefore in case of an inadvertent intravascular puncture, compression would be easy, and the consequences of bleeding would not be life-threatening. Joubert et al., in their meta-analysis, showed that the risk of bleeding in patients on antiplatelet and/or anticoagulants following peripheral nerve blocks is extremely low (0.64%-1.0%) (5). Compared with the consequences of giving GA to this patient, the risk of bleeding <1% was a safer option.

Conclusion

An ultrasound-guided axillary plexus block with ropivacaine allowed us to provide effective anesthesia for a patient with DMD with cardiomyopathy in CHF on the thrombolytic dose of heparin needing an emergency fasciotomy to avoid avoiding the thrombolytic dose the deleterious effects of GA in these patients.

Acknowledgment

None.

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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