Anaesthetic Management of a Case of Duchenne Muscular Dystrophy

Mohammad Ali, Fatema Johora

Abstract— Duchenne muscular dystrophy (DMD) is a x-linked recessive disorder characterized by progressive muscle weakness, wasting, sensory loss, and cardiomyopathy and respiratory failure in 2nd or 3rd decade of life. There is no established guideline for anaesthetic techniques in such patient. We report lengthening of tendo-achilis of both lower limbs in a 26 years old male who was diagnosed DMD since his childhood. Patient was found fit for anesthesia in preanaesthetic checkup. Operative procedure was done under subarachnoid block (SAB) with hyperbaric bupivacaine. Hemodynamic status of the patient was stable, and postoperative period was uneventful.

Index Terms— Anaesthetic challanges, Anaesthetic technique, Depolarizing muscle relax-ant, General anaesthesia, Inhalation anaesthetic agents, Myopathies, Total intraventous anaesthesia

1 Introduction

DUCHENNE muscular dystrophy is a x-linked recessive disorder with progressive muscle destruction that affects one in 3600 male infants because of mutations in the dystrophin gene. Onset is often in childhood, although some patients may present as adults. There is initially progressive weakness of girdle muscles, wasting and weakness are usually symmetrical without fasciculation or sensory loss, and tendon reflexes are present. Diagnosis may go unnoticed in the first years of life but is usually obvious by the 4th years, severe disability is typical by the age of 10 and may unable to walk after age 12. Cardiomyopathy and/ or respiratory failure may occur in 2nd or 3rd decade life [1], [2], [3].

Diagnosis is often clinical. CPK is grossly elevated (100-200 X normal), biopsy shows variations in muscle fiber size, necrosis, regeneration and replacement by fat, and on immunochemical staining, absence of dystrophin. Myopathic pattern is found in EMG. There is no curative treatment. Passive physiotherapy helps prevent contractures in the later stages. Steroids are sometimes used in treatment [1], [2], [3].

Prevalence of Duchenne muscular dystrophy in Bangladesh is unknown. An observation in eastern India (Bihar, West Bengal and Bangladesh) reports a higher incidence of the DMD cases in Muslims compared to Hindu population in that region [4]. We report a case of tendo-achilis lengthening under subarachonoid block (SAB) in a 26 years old male who was diagnosed as Duchenne muscular dystrophy from childhood.

2 Case Report

A 26 years old male, normotensive, nondiabetic was admitted in Asgar Ali Hospital Ltd. on 05/12/17 in the orthopedic department with the chief complaints of unable to walk due to straightening of both feet for last six months. He was diagnosed as a case of Duchenne muscular dystrophy since his childhood. The diagnosis was confirmed by muscle biopsy but the report was not provided by the patient. He has four brothers among them the elder one was died at the age of 30 years. The other brothers are physically healthy. The patient studied up to HSC, and now works as a drawing artist. On general examination, he was mildly anemic, pulse was 86 beats/ min, blood pressure was 110/ 70 mmHg, intellectually normal, highly cooperative, average built and generally weak. On systemic examination, muscle power of both upper limbs was normal though patient could not raise above head and of lower limbs was 3/5 with much emaciation due to atrophic changes. There was total straightening of both feet and patent could not walk or stand, he was bound to wheelchair. Other systemic findings were normal. During laboratory investigation, complete blood count (CBC), random blood sugar (RBS), serum creatinine, chest x-ray, ECG, echocardiography were found normal. Serum creatine kinase was 2348 U/L (normal 30-170 U/ L)

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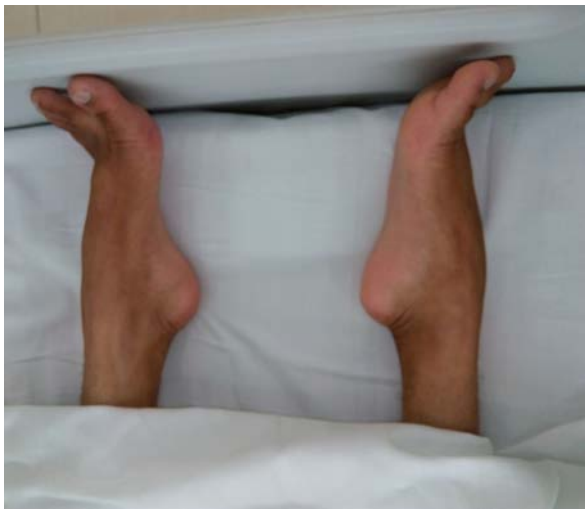

Figure 1: Lower limbs of the patient

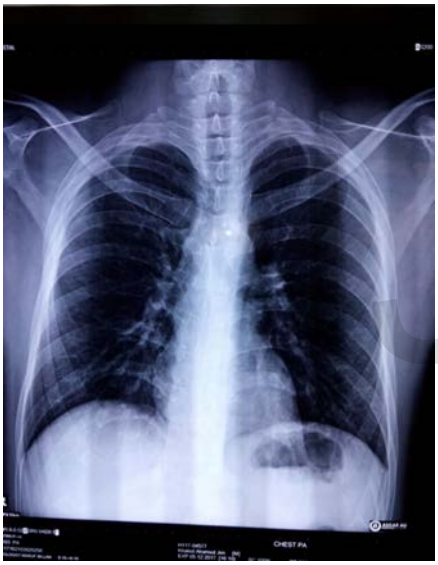

Figure 2: Chest x-ray of the patient

As the orthopedic surgeon decided to lengthen tendo-achiles of both lower limbs, the patient was taken to Operation Theater after proper evaluation and preparation. After I/V cannulation, patient was positioned for Sub-arachnoid block (SAB). SAB was given at the level of L3-L4 with 2 ml hyperbaric bupivacaine. Hartman solution was given during operation. After 10 minutes, patient was positioned prone for better surgical approach. Oxygen was delivered to the patient by nasal cannula the whole time during operation. There was no significant change in hemodynamic status of the patient. The operation took ninety minutes, and patient was able to move the toes after 120 minutes. Postoperative pain management was done by using paracetamol and ketorolac through I/V route. Postoperative period was uneventful.

3 Discussion

Patients of Duchenne muscular dystrophy represents a significant anaesthetic challenge as a result of their co-morbidities, in particular their lung function, and the volume of blood loss [5]. They are vulnerable to adverse effects and at high risk for death when they undergo procedures requiring general anesthesia and sedation. In these patients, exposure to depolarizing muscle relaxant, succinylcholine and halogenated inhalational anesthetic agents may be followed by atypical reactions and sudden cardiac arrest due to hyperkalemia resulting from massive rhabdomyolysis [3], [5], [6]. In myopathies, clinical decompensation has been reported during/ or after anaesthetic procedures with resulting hypoventilation, atelectasis, difficult extubation, dysphagia, arrhythmias and congestive heart failure [7], [8]. Patients with advanced stage of disease present with weakness of the oropharyngeal muscles including swallowing difficulties and possible elevated risk of aspiration. Studies did not find an increased risk of malignant hyperthermia susceptibility in patients with Duchenne muscular dystrophy (DMD) compared with the general population [9].

Normal preoperative ECG and echocardiogram findings do not exclude the possibility of severe cardiac complications in the perioperative period and cardiologist should be consulted in the preoperative period [10]. DMD patients have increased sensitivity for nondepolarizing neuromuscular blockers and, although they may be safely used with adequate monitoring. In general onset of neuromuscular block is delayed and duration is markedly prolonged. These effects are pronounced in advanced disease. Sedative drugs (benzodiazepines) can cause respiratory insufficiency [11].

There is no definite recommendation for either general or regional anaesthesia. General anaesthesia is contraindicated for non-essential procedures, especially in advanced stages of DMD. Total intravenous anaesthesia based anaesthesia is considered safest in these patients [11]. Nitrous oxide is generally considered safe. Regional anesthesia could be a good alternative, when possible, for minimally depressing respiratory and cardiovascular systems and requiring minor airway manipulation [3].There is reports of spinal, epidural and caudal anaesthesia without any complication. If muscular weakness is present and regional anaesthesia is planned, neurological consultation is helpful for juridical reasons. Opiates, propofol and local anaesthetics have been used without any complication. Patients may require a higher dose of propofol or opiates [12]. Researchers suggest a regional blockade as a safer alternative to general anaesthesia for the management of high risk DMD patients during orthopedic procedures [3], [13], [14].

4 Conclusion

In this case, surgical procedure was done under SAB and postoperative period was uneventful. Although there is no established guideline for surgical procedure and anaesthetic technique, experts recommend avoidance of general anaesthesia in avoidable cases. Patient with Duchenne Muscular Dystrophy needs careful evaluation during preanaesthetic check-up, individually and carefully anaesthetized on the basis of

medical judgments of anaesthesiologists and strict careful monitoring in postoperative period.

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