Brachial Plexus Block in a Patient with rare Genetic Disorder. Chronic Progressive External Opthalmoplegia - A Case Report

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Abstract

Background: Chronic Progressive External Opthalmoplegia(CPEO) is one of the rare mitochondrial gene disorders characterised by slow progressive paralysis of extra ocular muscles. Anesthetic drugs and techniques need to be modified for these patients. We present one such case

Case Report: A thirty-year-old female was posted for left wrist arthroscopy. Her previous reports revealed she was suffering from Chronic Progressive External Opthalmoplegia (CPEO). In view of possible complications with multidrug regimen it was decided to administer a dual mode ultrasound guided supraclavicular brachial plexus block. A total of 18ml of combined solution of xylocaine 1.5% and bupivacaine 0.5% were administered (Combined Supraclavicular and Axillary). The intraoperative period was uneventful throughout the surgical procedure that lasted for two hours. Neurological examination revealed a rapid onset motor loss (< 2 minutes) of the blocked limb without sparing of dermatomes. In the course of examination it was revealed that the patient complained of numbness of ipsilateral side of face. At follow up on the third day, it was discovered that the patient gave history of an unusual prolonged numbness of fingers which recovered completely after more than sixty hours and patient was discharged at this time.

Conclusion: An extended possibility of prolonged duration of the local anesthetic agent is possible after its application in nerve blocks. Regional nerve and plexus blocks are safer as compared to general anaesthesia in patients with mytochondrial myopathies

Keywords: Brachial Plexus Block, Chronic progressive external opthalmoplegia

Introduction

Chronic Progressive External Opthalmoplegia (CPEO) is one of the rare mitochondrial gene disorders characterized by slow progressive paralysis of extra ocular muscles [1]. Incidence of mitochondrial disorders is 9.2per 1ooooo adult population. Exact incidence of CPEO is not known but amounts to about half of total number of mitochondrial disorders [2]. Males and females are equally affected showing characteristic matrilineal pattern of inheritance.

Case History

A thirty year old female was posted for left wrist arthroscopy. Patient admitted history of initiation of disease process at her age of 19 years with feeling of exhaustion, drooping of eyelids and head-ache.A detailed clinical examination was not suggestive of involvement of any other neuromuscular or cardiac functions.Patient had an exclusive and isolated complaint of progressive symmetrical ex-traocular muscle paralysis. Her previous reports revealed she was suffering from Chronic progressive external opthalmoplegia (CPEO ()Figure 1). The mitochondrial disease was confirmed by Quadri-ceps muscle biopsy at her age of 24 years.

Preoperative period

Preoperative routine haematological investigations and chest X-ray were normal. ECG did reveal right bundle branch block pattern(Fig 1&2)

The general consensus in anesthetic management is to avoid non depolarizing muscle relaxants for fear of inadequate reversal and volatile anesthetic agents to avoid myocardial suppression and triggering of malignant hyperthermia. In view of possible complications with multidrug regimen it was decided to administer a supraclavicular brachial plexus block.

Anaesthesia technique

A dual mode, combined ultrasound and neurostimulation guided brachial plexus block by supraclavicular approach was planned for this purpose. A 10 ml of 0.5%sensorcaine and 10ml of 2%xylocaine with adrenaline was drawn in 20ml syringe. Under all aseptic precautions procedure was carried out with 22G insulated needle. Ultrasound modality was used to identify the needle tip at the corner pocket of supraclavicular area and neurostimulation to produce the evoked motor re-sponses of medial cord at 0.4MA.

On injection of 8ml of drug by supraclavicular route, as is routinely performed a proximal scan found the drug at the level of C6 root. Injection at supraclavicular area was abandoned at this point and it was decided to supplement with an axillary brachial plexus block. The axillary route was approached and 5 ml of drug was injected targeting the radial, and musculo-cutaneous nerves after eliciting the desired responses. A total of 13ml of combined solution of xylocaine with adrenaline

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Figure 1: Drooping eyelids of patient: Chronic Progressive External Ophthalmoplegia

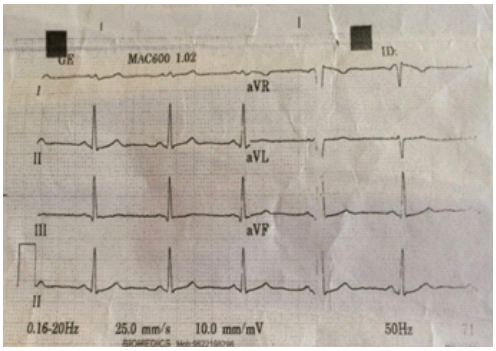

Figure 2: . ECG : Right Bundle branch Block pattern

2% and bupivacaine 0.5% were administered.

Intraoperative period

Patient was monitored by pulse oximetry, continuous electrocardiogram and NIBP. Patient was he-modynamically stable throughout the surgical procedure that lasted for two hours. Neurological examination revealed a rapid onset analgesia and anesthesia of the blocked limb with-out sparing of dermatomes. In the course of examination it was revealed that the patient complained of numbness of ipsilateral side of face. The numbness was spread across the dermatomes related to superficial cervical plexus upto the lower part of the ipsilateral ear, the ipsilateral aspect of the lip and the ipsilateral infraclavicular area and the proximal shoulder, which recovered towards the end of surgery and lasted for 2 hours.

Postoperative period

Patient was hemodynamically stable.
At followup, patient gave history of unusual prolonged numbness of fingers which recovered com-pletly after more than sixty hours

Discussion

Mitochondrial diseases are the result of DNA mutations of either nuclear or mitochondrial DNA. The respiratory chain is the essential final common pathway for aerobic metabolism, therefore tissues and organs that are dependent upon aerobic metabolism (central nervous system, heart) are preferentially affected in these disorders [3].

CPEO is a slowly progressive disorder. It may be isolated CPEO or associated finding of other mi-tochondrial disease such as Kerns Sayre syndrome [4,5,6].

Kearns-Sayre syndrome(KSS)first described by Kearns and Sayre in 1958 is the result of deletions in mitochondrial DNA (mtDNA) that cause a particular constellation of medical signs and symptoms. mtDNA is transmitted exclusively from the mother's ovum [7]. There is uneven distribution of dysfunctional mitochondria within each cell, and among different tissues of the body. This describes the term heteroplasmic which is characteristic of mitochondrial diseases including KSS. Thus two patients with an identical mutation in mtDNA can present with entirely different phenotypes and in turn different syndromes .

KSS is characterized by a triad of chronic progressive external ophthalmoplegia, pigmentary reti-nopathy, and at least one of the followings: cardiac conduction defects; cerebellar ataxia; or an elevated CSF protein level (＞100 mg/dl) that occurs before 20 years of age [8].

Atrioventricular block is the most common cardiac conduction defect which often progresses to third degree heart block. In the present case ,patient manifested with CPEO, RBBB, migraine, feeling of exhaustion and muscle weakness. We couldn't confirm diagnosis of KSS as reports of pigmentary retinopathy were not made available by the patient.

Anaesthetic implication

In KSS, with GA ,there is a risk of complete heart block, Congestive Cardiac Failure, postoperative respiratory dysfunction due to prolonged neuromuscular blockade and reduced respiratory drive with risk of malignant hyperthermia(unproven) [9]. Pre-medication with sedatives and opioids are avoided because the respiratory response to hypoxemia is impaired [10]. Propofol is safe (Shortand Young, 2003), but, in rare cases, propofol causes the so-called propofol infusion syndrome PRIS [11,12].

Despite this rarely occurring side effect, propofol has been also used without any complications in a large number of MID patients [13]. The volatile anaesthetics(haothane >iso=sevo) impair mitochondrial NADH oxidation (complex 1)and halothane also inhibits succinate oxidation(complex 2) thus exert negative inotropic effects [14]. In a patient with a variant of MID, however, anaesthesia, intro-duced with propofol and maintained with desflurane,

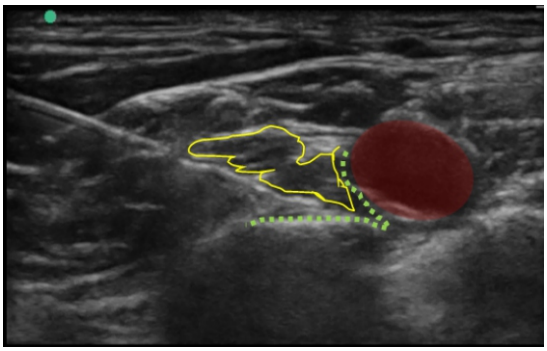

Figure 3: . Stimulating needle at the corner pocket
Green angle – Corner pocket (S C A & First Rib)
Yellow – Brachial Plexus
Red – Subclavian Artery (S C A)

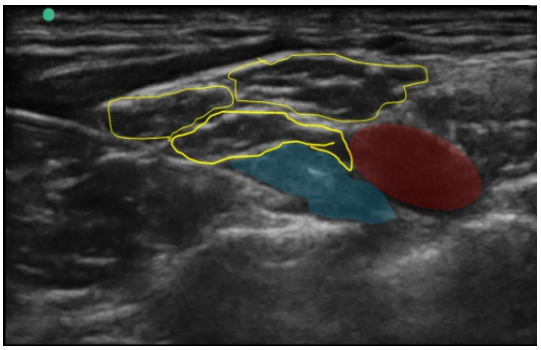

Figure 4: . Ultrasound Corner pocket
Supraclavicular injection 10ml 0.5% bupivacaine.
Red - S C A. Blue –L A spread. Yellow – Brachial Plexus displaced.

was followed by a fatal postoperative course, with unsuccessful weaning and death [15]. Volatile anesthetics also have a protective effect on mitochondria, which is responsible for anaesthetic preconditioning [16]. Amongst the non depolarizing muscle relaxants some resistance has been attributed to cisatracurium while an adverse reaction has been reported with rocuronium and atracurium. No adverse reactions have been reported with vecuronium [17].

As per literature local anesthetics have potential to worsen neuromuscular manifestations via direct effect on mitochondrial biogenetics [18,19,20]. Propacaine, cocaine, and tetracaine dissipate the mitochondrial membrane potential. Ropivacaine interferes with mitochondrial energy transduction, and bupivacaine directly affects mitochondrial function by depolarizing the mitochondrial membrane and oxidizing pyridine nucleotides. Mitochondrial toxicity was reported in one patient with KSS who received local anesthesia with articaine [21]. The prolonged duration of bupivacaine remains unexplained. Though there are reports of prolonged action of local anesthetics earlier [22,23,24,25]. Sixty hours is too long a duration for bupivacaine an-esthesia to regress.

As per published case report, general anaesthesia has been successfully administered in laproscopic cholecystectomy. However, keeping in view of the above mentioned complications in a patient of CPEO, regional anaesthesia was chosen as a safer alternative. In order to ensure if there was any neuromuscular residual effects, followup was done for 8 months which did not reveal any worsening of the existing disease.

Conclusion

Administration of any type of anaesthesia for surgical intervention in patients with coexisting dis-eases of genetic etiology like CPEO needs diligent approach. In present case recovery was uneventful but response to local anaesthesia was prolonged. Increased spread to level of trunks of brachial plexus was a procedural event which was well recognised under ultrasound guidance. The facial spread could be due to the maldistribution of the LA in the superficial cervical plexus area. Follow up of case for 8 months postoperatively did not reveal any deleterious effect of local anaesthetic on myopathic pathology.

Thus we can conclude, with words of caution that an extended possibility of prolonged duration in the post block period. Regional blocks are safer as compared to general anaesthesia in patients with a of mytochondrial myopathies.

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