Family Medicine and Primary Care: Open Access

Flampouraris V, et al. J Family Med Prim Care Open Acc 7: 252. www.doi.org/10.29011/2688-7460.100252

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Case Report



Laparoscopic Cholecystectomy under General Anaesthesia in a Patient with Steinert's Disease

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Citation: Flampouraris V, Ioannidis M, Grammenidis A, Nikolopoulou ZM, Fragkouli I, Kokolaki M (2024) Laparoscopic Cholecystectomy under General Anaesthesia in a Patient with Steinert's Disease. J Family Med Prim Care Open Acc 8: 252. DOI: 10.29011/2688-7460.100252

Received Date: 19 March, 2024; Accepted Date: 21 March, 2024; Published Date: 25 March, 2024

Abstract

We present a case of a man with Myotonic dystrophy Steinert type I, who underwent laparoscopic cholecystectomy under general anaesthesia and was discharged home on the next day.

Introduction

Myotonic Dystrophy often is abbreviated as «DM» in reference to its Greek name, dystrophia myotonica. Another name used occasionally for this disorder is Steinert disease after the German doctor who originally described the disorder.

Myotonic dystrophy type I or Steinert's disease is an autosomal dominant multisystem disease, which is characterized by consistent contracture of muscle following stimulation.

An abnormal nucleotide sequence on chromosome-19 causes prolonged stimulation of actin- myosin complex due to a larger sodium current, causing delayed relaxation of contracted muscle.

There are weakness and wasting of voluntary muscles in the face, neck and legs in type 1 myotonic dystrophy. Muscles between the ribs and those of diaphragm, which moves up and down to allow inhalation and exhalation of air, also can be weakened.

Case Report

A 48- years old male patient (170 cm, 82 kg, BMI: 28.37), with diagnosis of MD was scheduled for laparoscopic cholecystectomy because of 3 attacks of acute cholecystitis in last 6 months.

At the properative examination, the patient was alert and oriented. He was not able to walk unassisted and attend to his own bodily needs assistance (Ataxia).

On examination, patient was noted to have proximal and distal extremity weakness. ECG showed sinus rhythm 75 per minute.

We choose general anaesthesia.

We put in 18 G and 20 G intravenous line in situ in the right arm. We used measuring with radial arterial line (in the left arm) BP, SPO2, BIS, ECG, N.M.T train of four, ABGS and urine output.

Rapid sequence induction was accomplished by giving: Dexamethasone 8 mg, fentanyl 50 μ g, propofol (2-6 isopropylophenol) 1% 200 mg, rocuronium 20 mg (low dose) IV. Intubation of trachea with an 8.0 size endotracheal tube was facilitated with MAC 4.

Maintenance of anaesthesia with Propofol 1% 50-100 μ g/kg/min, and Remifentanyl 0.05-0.1 μ g/kg/min.

Ondasetron 4 mg, fentanyl 250 μ g, paracetamol 1gr, tramadol 100 mg, omeprezol 40 mg and mefoxil 1gr given IV. We used warm intravenous fluids R. lactate to maintain normothermia.

NMT monitoring used on orbicularis oculi muscle, showed no twitches on train of four for the rest of operation.

Sugammadex 200 mg IV was given at the end of procedure.

Preoperative ABGS

FiO2 0.21

PH 7.39, PO2 63 mmHg, PCO2 52 mmHg , HCO3 28.7, BE 5.0, SPO2 92%, Lac 1.5

Operative ABGS

FiO2 0.4

Volume 8; Issue 01

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PH 7.50, PO2 99, PCO2 38, HCO3 29.7, BE 6.0, SPO2 100%, Lac 0.9. (VT: 475 ml, f: 13/1', PEEP 5, I:E ½)

FiO2 O.4

PH 7.37, PO2 86, PCO2 49, HCO3 26.2, BE 2.1, SPO2 100%, Lac 1.0(VT: 475ml, f: 14 /1', PEEP 5, I: E ½)

When the patient was alert (fully awake) and breathing spontaneously, train of four 4/4 the trachea extubated.

ABGS

PH 7.31, PCO2 59, PO2 70, Lac 1.0, BE 2.2, SPO2 92%, HCO3 29.7.

M. Venturi O.35.

After that, we used BiPAP for 20 minutes.

At least M. Venturi FiO2 O.31 ABGS:

PH 7.37, PO2 86, PCO2 49, Lac 1.2, HCO3 26.2, BE 2.1, SPO2 96%.

In the PACU, the patient was kept warm with a forced air blanket, hemodynamically stable, VAS 0/10.

Discussion

We used rapid sequence induction because in these patients exists high risk of aspiration.

We choose total IV anaesthesia, avoids volatile anesthetics, which often cause myotonic crisis and shivering.

Rocuronium is safe as muscle relaxant.

Sugammadex is the best solution because neostigmine and fysostigmine have been found to cause incomplete reserval in patients with DM.

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