Anesthetic Considerations of a Patient with Nemaline Myopathy: A Case Report

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Introduction

Nemaline Myopathy (NM) is a rare congenital myopathy defined by generalized muscle weakness and the presence of nemaline bodies seen on muscle biopsy. It is associated with respiratory complications, facial dysmorphism and skeletal deformities.

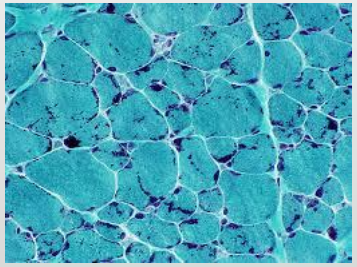
Case Report

A 17 year old female with history of NM presented for tenotomy, tendon lengthening, and gastrocnemius release. She has a history of general anesthesia at age three resulting in possible MH. Patient is tracheostomy and ventilator dependent, on chronic pyridostigmine for congenital myasthenic syndrome, and has history of chronic respiratory failure, cardiomyopathy, scoliosis and developmental delays. She also suffers from severe anxiety disorder including needle phobia.

MH precautions were taken which included flushing the machine and avoidance of all triggering agents. Midazolam and ketamine premedications were given through the gastrostomy tube thirty minutes prior to the operation. She was on home transport ventilator with back-up rate for apnea. Once the patient was in the operating room, we placed her on the anesthesia ventilator, gave 50% nitrous oxide while obtaining a peripheral IV. We then began total intravenous anesthesia using propofol drip and dexmedetomidine bolus. We avoided using succinylcholine, depolarizing muscle relaxants, and volatile anesthetics. We also avoided long-acting narcotics due to her myathenic syndrome. Pain control was achieved with acetaminophen, toradol, and fentanyl. Intraoperative hemodynamics and temperature were stable.

Discussion

We took MH precautions due to the mother reporting a history of fever from anesthesia when child was 3 years old. No case of MH with NM has been reported in the literature, but the disease is associated with other congenital myopathies including central core disease with a predisposition to MH. Pulmonary complications and difficulty with intubation are the more commonly reported issues of NM and anesthesia. NM weakness affects respiratory muscles, and this can lead to respiratory dysfunction including postoperative respiratory infection and prolonged ventilation. Facial abnormalities like a high-arched palate and micrognathia can make tracheal intubation challenging. We were able to judiciously premedicate since the patient was ventilator dependent. This allowed greater ease in obtaining our IV and administering a safe TIVA.



Gomori trichrome staining of muscle-biopsy sections. The deep-purple staining of namaline bodies indicated by arrows.

Conclusion

This case exhibited a complicated patient with an exceedingly rare muscular disease and multiple conflicting pathologies. This case required creative use of premedication for induction followed by a well performed TIVA that was conducive in preventing further muscle weakness due to her myasthenic syndrome.

References

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