CASE REPORT

An unusual presentation of postoperative malignant hyperthermia

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ABSTRACT

An anesthesiologist may have only one odd occasional encounter with malignant hyperthermia throughout his or her clinical career, but it is always a reminder of our professional inadequacies, may it be clinical management, or availability of essential drugs or equipment. Malignant hyperthermia strikes when one least expects, as it happened in a child in postop recovery in a cleft camp. Quick decision and ready availability of dantrolene controlled the syndrome.

Key words: Malignant hyperthermia; Intraoperative Complications; Dantrolene; Body Temperature; Postoperative Complications; Ryanodine Receptor Calcium Release Channel; RyR1

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INTRODUCTION

Malignant hyperthermia (MH) is a genetically determined life-threatening clinical syndrome of hypermetabolism involving the skeletal muscle. It is triggered in susceptible people after being exposed to certain inhalational anesthetic agents and the muscle relaxant suxamethonium. Clinical picture ranges from muscle rigidity to raging temperature with variation in time of onset. We present a case of six months old child who underwent cleft lip surgery under general anesthesia (GA). The course of anesthesia was uneventful but the baby's body temperature rapidly spiked during recovery. The temperature was brought under control with inj. dantroline. We could not find similar case report in the published literature.

CASE REPORT

A six month old child of 6 kg presented for repair of cleft lip in our recently concluded cleft lip surgical camp at Gujrat (Pakistan). Apart from having a syndromic look, child was fairly fit and healthy. There was no cardiac or congenital anomaly or history of any other problem; chest was clear, temperature was normal and preoperative investigations including hemoglobin and white cell count were all within normal limits. The child had had cleft surgery under GA, two months ago in the same hospital, which remained uneventful.

The baby was placed nil by mouth for four hours. In the operating room (OR), basic monitoring (ECG, pulse oximeter and precordial stethoscope) were applied before induction of GA was done with 6-8 % sevoflurane in oxygen and inj. atracurium 0.5 mg/kg. Child was intubated with size 3.5 endotracheal tube (ETT) and position of the tube confirmed with bilateral breath sounds and capnography. Child was manually ventilated and anesthesia maintained with sevoflurane in oxygen and nitrous oxide using Ayer's T piece. Throughout the intraoperative period her SpO_2 remained 99%, heart rate at 130 -140/min and $EtCO_2$ remained at 3.5-4.5 Kpa. Surgery finished in 45 min with minimal blood loss. As per local protocol inj. paracetamol 90 mg and tramadol 1 mg/kg was given for analgesia. Child was extubated and transferred to recovery after an uneventful anesthesia.

In the recovery, child's heart rate and temperature started to rise steadily. The high temperature did not settle with conventional measures like cold sponging and intravenous fluids and continued to rise steeply. Within 45 min her temperature had increased to 104ª F, and heart rate increased to 180 / min. In the next 15 min her temperature was touching 105.5ª F with heart rate at 225 / min and respiratory rate to 40 breaths per min. Her

Table 1: Pre- & post-dantrolene vital signs

Time	Temperature ºF	Heart Rate (per min)	SpO_2 (%)	Respiratory rate (per min)
10:20	100	140	98	30
10:25	102	154	96	35
10:30	103	166	95	35
10:35	104	180	90	38
10:40	105	188	92	40
10:45	105.5	225	89	40
Dantrolene 1 mg/kg IV				
11:00	104	190	90	40
11:30	101	165	95	36
11:45	100	150	96	34
12:15	102	170	94	36
Dantrolene 1 mg/kg repeated				
12:45	99	140	97	34

condition was getting worse by the minutes with no sign of improvement. At this point our working diagnosis was either an acute infective episode (viral/bacterial/ malarial) or MH.

We had limited resources in the free cleft camp, with no pediatric ventilator or pediatric defibrillator and had only limited blood tests available. There was no question of availability of inj. dantrolene in this hospital. Fortunately we had packed few ampules of dantrolene with us, while embarking for our journey. The drug was donated by one of the private hospitals in UK. At 105.5ª F we decided to use it. The drug was prepared by dissolving 60 ml of water in dantrolene vial to give a dilution of 0.3 mg/ml. An initial dose of 1 mg/kg was administered intravenously.

Whatever form of hyperthermia it was, it responded to dantrolene immediately. Temperature and heart rate started to decline steadily (as shown in Table 1). We repeated dantrolene after one hour when temperature began to rise again and it again responded favorably. The only blood test available was full blood count. It showed Hb of 11 g/dl, WBC count of 10,000 and platelets at 46000. Child stayed in the recovery area overnight with continuous monitoring of temperature, heart rate and oxygen saturation. Only side effect noticed with dantrolene was drowsiness which disappeared the following day as child made full recovery.

DISCUSSION

MH is an autosomal dominant disorder of pharmacogenetic origin in which the suspected individual exhibits hypermetabolism after exposure to one or more triggering agents, including potent inhalational agents e.g. halothane, isoflurane, sevoflurane etc. Succinylcholine is another agent contributing to MH although its individual role is controversial.[1]

MH occurs as a result of a disturbance in calcium homeostasis due to genetic defects responsible for excitation contraction coupling of dihydropyridine ryanodine receptor complex. It can be a life threatening emergency if immediate removal of triggering agent and prompt management does not take place. Incidence varies from 1:50000 to 1:150000 of general anesthetics administered.[2] Previously, the mortality rate was 70-80%, which has now decreased to 3-4% due to awareness and monitoring and availability of dantrolene.

Dantrolene reduces free intracellular calcium by binding to ryanodine receptor thus depresses skeletal muscle excitation- contraction coupling. All the anesthetic professional bodies around the world advise that an adequate supply of dantrolene must be available wherever GA is administered. Dantrolene should be available within 10 min of the decision to treat MH.

MH presentations vary from case to case. Previous history of uneventful anesthesia does not preclude MH despite receiving all triggering agents. Its magnitude of severity also varies. The time of onset can range from just after introduction of a triggering agent to 40 minutes after discontinuation of anesthesia. Carpenter et al in their study have demonstrated that time interval between induction and presentation plays a role in severity of MH.[3]

Larash grading system based on various clinical signs and tests[4] is widely used for diagnosis of MH. Greater the score, greater is the chance of MH. A score of greater than 50 is almost 100% diagnostic. Despite not having access to advanced blood testing, our child scored 30 in Larash grading which made it more probable to be MH. Postoperative MH occurred only in 1.9% of the cases and classic presentation of MH was rarely found such as muscle rigidity, hyperthermia, tachycardia and it could present with delayed rhabdomyolysis with latency period ranged between 0 to 40 min.[5,6] Presentation in our case was within this time period.

Our patient did not have muscle rigidity and we had no access to CK testing. Nelson et al demonstrated differences in clinical presentation in pediatric patients. They found that youngest age group patients were less

likely to develop muscle rigidity. The most common clinical presentation were tachycardia (74.3%), tachypnea (77.1%) increase in temperature (60%) and skin mottling (14.3%). Muscular rigidity was found only in 17.1% and in those patients who were given succinylcholine.[7] Our child had all those signs. MH as thought earlier can have hidden presentation, not appearing intraoperatively and can be fulminant if not picked early. Care should be taken with suspected syndromic patients.[8] In our case the child had a syndromic look. Retrospectively, we think it could be King Denborogh Syndrome (KDS) which is a congenital myopathy with musculoskeletal abnormalities, and definitely associated with susceptibility to MH.[9]

This case raises the issue of public and private hospitals in smaller cities of Pakistan being ill equipped; where an arterial blood gas analysis and creatinine kinase testing is only a dream. This case also highlights the challenges of working in a resource poor country where there is no availability of life saving drugs like dantrolene.[10] A quick survey of the multiple tertiary care hospitals in Pakistan revealed that dantrolene is non-existent in Pakistan. This should be an area to reflect upon, not only by the anesthetist community in Pakistan but also by the big international organisations like WFSA. Steps should be taken to make life saving drugs like dantrolene, pooled together in at least one place in a city so that there is no unnecessary loss of life.

Conflict of interest: Nil

Authors' contribution: All the authors took part in the conduct of case and manuscript preparation.

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