Malignant Hyperthermia with Disseminated Intravascular Coagulation during Congenital Ptosis Correction: A Case Report

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Abstract The case of a 10-year old Caucasian boy operated for correction of congenital bilateral ptosis is described. The picture of a malignant hyperthermia complicated with disseminated intravascular coagulation was seen during the surgery, almost twenty minutes after inhalational volatile anesthesia started. The patient was pronounced dead after the third episode of cardiac arrest that did not respond to electrical conversion. Microscopic and macroscopic findings of the autopsy are described as well, with diffuse bleeding and extravasation in almost all vital organs and body cavities. A discussion on the triggering factors and neuromuscular disorders that are particularly prone to get complicated with imputed anesthetic agents is made, with special referral to similar cases found in the literature, since such a complication was initially reported.

Keywords: halothane, congenital ptosis, malignant hyperthermia, disseminated intravascular coagulation, dantrolene


1. Introduction

Malignant hyperthermia (MH) is a potentially lethal complication, considered a rare disorder that is elicited mainly during anesthesia in susceptible individuals. Described initially in 1960 from Denborough, such a complication has been continuously under scrutiny, with several attempts to produce reliable tests that might forecast and prevent such an event [1,2,3]. Different nosologies, mainly falling into the neuromuscular family of disorders and several eliciting drugs have been imputed (Table 1). Recent and meticulous research has substantially reformulated the theoretical basis for such a complication, actually considered as a disease of calcium-regulating proteins [4]. Based on the theories of an abnormal skeletal calcium release during the cascade of the events, confirmation of the diagnosis of MH actually is reliably made through ryanodine receptor gene sequencing [5,6,7]. The use of dantrolene has considerably influenced the prognosis of such a disorder, but other appropriate symptomatic measures have to be honored, with discontinuation of offending drug, treatment of acidosis, electrolyte disbalance and heart rhythm disorders, among others [8]. Worth mentioning is the fact that MH during anesthesia represents a relatively rare event, with an incidence estimated from a minimum of 1:10000 interventions from some sources, and a maximum of 1:220000 in others [9,10]. There is however a general accord that the disorder is more frequent during anesthesia in children [11].

2. Case Report

We report a fatal case of MH complicated with disseminated intravascular coagulation (DIC) during a cosmetic intervention for bilateral congenital ptosis, in a 10-year old Caucasian boy.

The anesthesia was induced with propofol and fentanyl; vecuronium was given as a myorelaxant and anesthesia was maintained with inhalational-volatile agents, formed by a mixture of halothane, oxygen and air.

Less than twenty minutes after initiation of volatile anesthesia, profound skin erythema and hyperthermia were installed, with the body temperature reaching 41 Celsius degrees. Tachycardia (130 heart beats per minute) and tachypnea (35 respirations per minute) forced the anesthesiologist to take adequate measures, and surgical procedures were immediately stopped. Perfusions and antipyretics were given, but the body temperature did not fall below 41 Celsius degrees. Profuse hemorrhage in the suture areas opened during the initiation of surgery was seen and treated through local compression.
The patient became mydriatic fifty five minutes after initiation of volatile anesthesia and suffered twice a cardiac arrest. Electric cardioconversion was applied successfully with returning of sinus rhythm although tachycardia persisted; the third cardiac arrest followed few minutes after the first hour of volatile anesthesia was completed and resulted fatal. The patient was pronounced dead half than an hour later and the autopsy was performed in a nearby forensic facility. Diffuse hemorrhage was seen inside intra-abdominal and intra-thoracic cavity, with macroscopic changes due to red blood cells extravasation and liquid leakage (Figure 1 and Figure 2).

Figure 1. Macroscopic view of the lungs, with diffuse hemorrhagic changes

Figure 2. Macroscopic view of the stomach, with massive bleeding

Figure 3: Microscopic view of the lungs, with interstitial hemorrhage (Hematoxylin-Eosin, 160X)

Figure 4. Microscopic view of the cerebellum, with interstitial hemorrhage (Hematoxylin-Eosin, 160X)

The diffuse character of the hemorrhage suggested the clinical picture of an intravascular disseminated coagulation, a situation that might complicate malignant hyperthermia. Microscopic changes encountered confirmed the diffuse bleeding and red blood cells extravasation; Figure 3 and Figure 4 illustrate changes in two of the organs sliced.

3. Discussion

Several drugs have been accused and considered as unsafe in patients susceptible to malignant hyperthermia; Table 1 summarizes some of those drugs.

Table 1. Drugs contraindicated in patients susceptible to malignant hyperthermia

| 1. Volatile anesthetics       | a. Halothane        |
| 2. Myorelaxants              | a. Succinylcholine  |

(Adapted from 11)

The role of calcium channel blockers is controversial; the over-mentioned author considers these drugs as unsafe, whereas other sources grant therapeutic potency to diltiazem and nifedipine (two well-known calcium channel blockers) regarding contractures caused from halothane during the symptomatology of halothane-induced malignant hyperthermia [12]. Several case reports of malignant hyperthermia during anesthesia with volatile agents are suggesting different approaches when the condition is unexpectedly presented. Some authors suggest that malignant hyperthermia might resolve merely through the discontinuation of the offending drug [13]. However, dantrolene usage is generally considered as necessary and efficacious, albeit other drugs such as procainamide, sodium bicarbonate, furosemide etc. have a certain utility and should be available in the operating room [11].

Dantrolene sodium is not licensed and is not available in Albania till the date the present manuscript was written. The fact is that the drug has a very narrow pharmacological scope of usage and even in larger countries is considered as an ‘orphan drug’ [14]. Difficulties regarding the overall availability of the drug in the operative theatres have been formulated elsewhere,
with cases that never received dantrolene sodium although the necessity was clear [15].

Table 2 summarizes some of the major disorders that are associated with the presentation of malignant hyperthermia, and thus considered as major risk factors when anesthesiologists should decide over the usage of certain unsafe drugs, such as those listed in the Table 1.

### Table 2. Disorders considered as at risk for presenting malignant hyperthermia

1. Asymptomatic familial increase of CPK level
2. Central-core disease
3. Duchenne muscular dystrophy
4. Myotonia
5. Schwartz-Jampel syndrome
6. Congenital muscular dystrophies

(Adapted from 16-18)

As summarized in the table above, patients highly risked for malignant hyperthermia are all those suffering from neuromuscular disorders; drugs considered as safe for anesthesia in them have been identified, with barbiturates, ketamine, fentanyl and propofol being on the top of the list. The patient we described in the present case had a congenital bilateral ptosis, but no signs of muscular suffering elsewhere. However, we would suggest in view of this fatal complication, a tight control of CPK values even to patients with ‘benign’ ptosis, since even the origin of the latter, is a matter of wide controversy. In fact, myopathies might present with bilateral congenital ptosis, till the complete symptomatology of the disease will become clear [19]. The fact that neuromuscular complications (including malignant hyperthermia) are extremely rare, and that congenital ptosis correction is technically non problematic, explains the high level of surgical success reported, parallel to almost a complete lack of such severe complications [20].

### References