Concerns for Non-operating Room Anesthesia in a Child with Dravet Syndrome : A Case Report

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ABSTRACT

Introduction: SMEI - Severe myoclonic epilepsy of infancy which later came to be known as Dravet syndrome due to its variable symptomatology, is a rare epileptic syndrome. Seizures are refractory to anti-epileptics and the syndrome is associated with cognitive impairment. Various studies have shown abnormal body temperature regulation as the underlying cause for seizures along with other triggers like stress and fever.

Case report: We present a case of a seven year female patient undergoing diagnostic MRI and the challenges faced during the procedural sedation. The procedure was uneventful, but there was a slight difficulty in iv cannulation due to the agitated child and hyperthermia during the procedure. Both of the problems were managed in a feasible way due to NORA.

Conclusion: Patients with this syndrome pose unique challenges for anesthetic management, especially in a resource limited non-operating room environment as compared to other pediatric patients. By the observations made in our case, we conclude that the goals of NORA in a case of dravet syndrome are:

1. Adequate pre-anaesthetic evaluation including anti-epilepic drug history and to continue it on the day of procedure.
2. To maintain normothermia and,
3. To prevent pre or post-procedure agitation.

Keywords: Non operating Room Anesthesia, Dravet Syndrome, SMEI, Severe Myoclonic Epilepsy of Infancy, Seizures.

INTRODUCTION

Dravet syndrome, previously called Severe myoclonic epilepsy of infancy is a partial epileptic encephalopathy syndrome associated with cognitive impairment with increasing age.[1] It was first described in 1978, France. It was renamed to a syndrome due to variability of the symptomatology and the absence of myoclonic components sometimes. Seizures are often refractory to anti-epileptics, therefore are challenging to treat.[1] These patients often undergo various minor diagnostic procedures like feeding tube insertion, diagnostic MRI, catheterization which need sedation and anesthesia management, which can be challenging in a non-operating room environment. We document the anesthetic challenges faced during MRI in a case of dravet syndrome.

CASE PRESENTATION

A 7 year old female patient diagnosed with Dravet syndrome was scheduled for diagnostic MRI in view of increased episodes of seizures since two to three years. Her medical history included seizures from the age of 9 months, initially started as febrile seizures later increased to 2 to 3 episodes a year. Recently, the number of episodes have increased to six to eight episodes a year, often associated with status epilepticus. Mental slowing had been observed by the parents since 1 to 2 years of age. The patient is on anti-epileptic sodium valproate and topiramate.

In the pre-procedure room, the patient was made to sit comfortably on the mother's lap, NBM was confirmed. The patient was noted to have agitation on visit by the anesthesia care takers. Hence, the patient was allowed to relax. With positive reinforcement, anesthesiologists were able to reduce the agitation, which is a major seizure inducing trigger in Dravet syndrome.[2] The vitals were recorded and were found to be within normal limits. The patient's body (axillary) temperature was 98.2 degree F. The temperature inside the MRI room was 68 degree F.

The intravenous access was secured with slight difficulty and midazolam 2.5 mg given intravenously. The calm and co-operative patient was then allowed to undergo an MRI scan. A bolus dose of propofol 20 mg was given to achieve adequate sedation intra-procedure. Duration of the MRI was 20 minutes. During the procedure, the HR, saturation were continuously monitored and temperature was monitored intermittently. MRI compatible airway equipment and resuscitation devices were kept on standby.

Although the procedure was uneventful, the patient's temperature momentarily spiked from baseline to 99 degree fahrenheit during the procedure. The reason for increase in body temperature was suspected to be due to cotton wrapping which was done to prevent hypothermia in the extremely cold MRI room which was maintained at 68 degree F (20 degree C). The wrapping was removed and cold saline bags were used to decrease the temperature from the body surface. Post-procedure, the patient was calm, vitals were stable. The

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patient was observed for 2 hours and then sent to the parent department for further management.

DISCUSSION

SMEI - Severe myoclonic epilepsy of infancy which later came to be known as Dravet syndrome due to its variable symptomatology, is a rare epileptic syndrome. Mutations in the SCN1A gene responsible for proper functioning of voltage gated Na channels are affected. Because of this defect the seizures are refractory to commonly used anticonvulsants. Inappropriate anti-epileptics like carbamazepine, oxcarbazepine administered initially when the diagnosis is in doubt can increase the seizure activity, therefore Levetiracetam, sodium valproate, Potassium bromide are found to be effective in treating Dravet syndrome.[1,2,5]

Because of the refractory nature, the seizures are poorly controlled in Dravet syndrome. Hence, preventing perioperative seizures is an important concern in anesthetic management. The following are the potential problems that can be faced in dravet syndrome during anesthesia:

Overheating

Temperatures even slightly higher than 37 degree celsius can sometimes trigger convulsions in Dravet syndrome.[2] Warm, humid weather, anti-hypothermia measures usually taken for general pediatric anesthetics like, increasing OR temperatures, forced air warmers, warm iv fluids, cotton wraps can be detrimental in Dravet syndrome. In our case, this trigger was identified and managed in a feasible way (cold saline bags) since it was a resource limited non- operating room environment. Irrespective of the circumstances, maintaining normothermia must be one of the goals of anesthetic management.

Stressful triggers

Agitation due to hospital environments, separation from the parent, equipment and MRI related Claustrophobia can be difficult to tackle for an anesthesia provider. Patients of Dravet syndrome have cognitive impairment, therefore can unusually react to these triggers, increasing the difficulty in pre-procedure venous access and sedation. Therefore, building a rapport with the patient and providing a comfortable environment can decrease the risk of perioperative seizures. In our case, there was a slight difficulty in placement of IV access due to the agitated patient, but was managed successfully. The patient was calm and compliant after sedation.

Abnormal temperature regulation

Dravet syndrome patients have abnormal temperature regulation due to the effects of anticonvulsant medications on hypothalamus, this can be exaggerated due to general anesthesia.[3] In a non-operating room environment, access to good temperature monitoring devices and cooling or heating equipment is not possible. Therefore, alternative modalities of temperature control are necessary. In our case, a fiberoptic skin temperature probe was used for monitoring and cold saline bags were used for physical cooling after hyperthermia was identified. Care must be taken that the patient is not deliberately made susceptible to hypothermia by overzealous measures. Prevention of hypothermia and hyperthermia simultaneously can be challenging especially in pediatric patients due to above mentioned reasons in dravet syndrome.

Fever and malignant hyperthermia

As these patients may have low immunity and FTT (failure to thrive), infection and fever are other concerns which may increase the risk of seizures. Risk benefit ratio of postponing the procedure till the URI, LRI, GE resolves should be weighed cautiously. Appropriate antipyretics should be given peri-operatively if necessary. With the use of sevoflurane and succinylcholine, the risk of malignant hyperthermia should always be kept in mind.

CONCLUSION

Although the incidence of Dravet syndrome is rare, 1:20000 to 1:40000 births, it is possible that the syndrome is underdiagnosed due to its varied symptomatology. Therefore, any pediatric patient with a history of febrile seizures should be cautiously considered for anesthesia. Minor procedures like MRI should be carried out with a proper anesthetic plan. By the observations made in our case, we conclude that the goals of NORA in a case of dravet syndrome are:

1. Adequate pre-anaesthetic evaluation including anti-epilepic drug history and to continue it on the day of procedure.
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