Anesthetic Management Of A Child With Sickle Cell Disease For Total Spine MRI in BrainSuite®

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Abstract

BrainSuite® (BrainSuite) is an advance in operating neurosurgical operating room.[1] It contains Intra-operative MRI (iMRI) with limited magnetic field effect which allow surgeon and anesthesiologists to work inside the room and away from the magnetic field of iMRI. It has all the luxury of an operating room for surgery and all the luxury of imaging and iMRI scanning. However, the problem for a child affected by sickle cells disease and spine osteomyelitis was that theprocedure got declined radiology MRI for fear of cold environment effect on the sickling process and the lack of temperature monitoring in that building.
The investigation was done in iMRI of BrainSuite safely under warming devices and full cardio- vascular, respiratory and temperature monitoring.

INTRODUCTION

The safety of modern Anesthesia increased the demand on its services. This widened the scope from therapeutic surgical intervention to diagnostic procedures in many fields in medicine. This includes children going for magnetic resonance imaging (MRI). MRI brain scanning in Sickle Cell Disease (SCD) has been reported in cases suffering from cerebral vascular accident (CVA).

Anesthesia for children going for MRI is important demand in order to have control of the child during the procedure. This can be accomplished using anesthetic machines (Aestira 5 MRI Datex - Ohmeda) and monitoring devices (Millennia 3155 MVS Invivo) compatible with high powered magnetic field. We report an unusual case of SCD suffering from osteomyelitis of spinal vertebra. It was declined the MRI in radiological department because of fear of hypothermia inside MRI room which have detrimental sequences on sickle red blood cells. The attending surgeon indicated the importance of this study in order to help him in deciding his surgical plan.

The dilemma was solved by scheduling this patient to be done in BrainSuite® (BrainSuite)'s Intra-operative MRI (iMRI) at KFMC on the ground of the availability of worming device (Ari Zant Healthcare Inc.-USA) and monitoring of temperature during surgery. The issue was not totally ideal since there were need to interrupt the heating during the iMRI session. BrainSuite is an advance in operating neurosurgical operating room.[1] It contains iMRI with limited magnetic field effect which allow surgeon and anesthesiologists to work inside the room and away from the magnetic field of iMRI. It has all the luxury of operating room for surgery and all the luxury of imaging and iMRI scanning. This in advance made it in the authors mind a problem solving for MRI investigation of child affected by sickle cells disease and spine osteomyelitis , Who was declined anesthesia in radiology MRI for fear of cold environment effect on the sickling process and the lack of temperature monitoring in that building.

The investigation was done in iMRI of BrainSuite safely under worming devices and full cardio- vascular, respiratory and temperature monitoring.

Anesthesia team took the challenges and the patient was safely anesthetized and MRI revealed osteomyelitis of spinal vertebra.

CASE REPORT

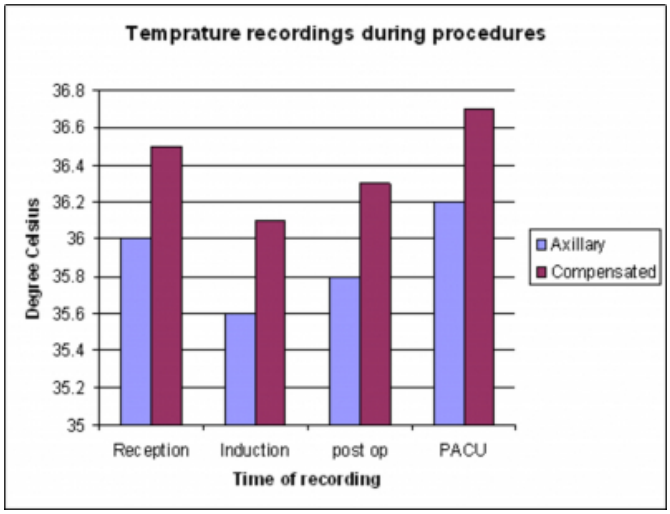
A 9 years old female child weighing 14.6 Kg was admitted to KFMC Pediatric Hospital on 31 October 2007 due to uncontrollable fever and back pain, she was a known case of SCD. Computerized Tomography Scan (CTS) was done without anesthesia demonstrating loss of bony substance of the body LV2 with endplate irregularities and loss of the disc spaces above and below consistent with deossification (due to marrow hyperplasia) infection or infarction as a sequel of

SCD. The neurosurgeon requested MRI spine to role out the diagnosis of osteomyelitis of lumber vertebra. In preoperative evaluation visit the child was in good general condition and without any evidence of end-organ damage, afebrile, stable vital singes, neurological examination free, not distressed, good bilateral air entry without added sounds, S1 and S2 regular no murmurs, lab results: anemic Hb 8.08 mg/dl, Ht 25.6 %, WBC 13.4, Platelets 192, Na 136, K 3.6, Creat. 35 (53-88), Urea 2.4 (2.5-6.4), Alkaline phosphates 249 (50-136), Bilur. T. 58.3 (0-17.1), Bilur. D. 27.4 (0-5), ALT (SGPT) 84 (30-65), PT 14.5, INR 1.19, PTT 36.4, Hb A 81.3%, Hb A2 2%, HbS 16.7%. the anesthesia order was NPO 6 hrs prior procedure and iv fluid D1/4S 50 mls/h starting from fating time. The anesthesiologist started in maintaining the temperature immediately after the child arrived to operating theatre's reception area by Bair Hugger on 38°C for the duration of one hour. Then the child was transferred to induction room in BrainSuite and also the anesthesiologist restarted warming by using Bair Hugger on 43°C. The child was induced by inhalation anesthesia by using mixture of 100% O_2 and 8 vol. % sevoflurane. When the child was deeply anesthetized LMA size (2) was inserted smoothly. Then the child was transferred to BrainSuite and completely (including the head) covered by three warmed blanket, one under and two above her body. During procedure the ECG, heart rate, blood pressure and $EtCO_2$ was monitored and kept with acceptable range. The procedure took 62 minutes. Anesthesia was maintained by a mixture of 100% O_2 and 3 vol % sevoflurane. The patient was maintained spontaneous breathing all through the procedure., Warm intravenous (iv) Lactated Ringer's solution during procedure at a rate of 75 ml/h After that the child was transferred again to induction room, warming by Bair Hugger restarted by 43°C, 100% O_2.

LMA removed smoothly then the child was positioned in lateral position spontaneous breathing O_2 enriched air via face mask, the child transferred finally to PACU after full recovery.

MRI finding: consistent with bony Ostiomylitis secondary to SCD in lumber spine. She was but on a course of intravenous suitable antibiotic were she showed excellent response without the need for any surgical intervention

Figure 1



DISCUSSION

The MRI procedure for this child was cancelled two times in the radiology's MRI department due to unavailability of warming facility for the child during imaging session. The ambient temperature is cool for child who may sickle if he gets hypothermic. This delay led to increased child suffering, as he stayed undiagnosed and has long hospital stay.

What were the considerations governing anesthesia management?

THE FIRST CONSIDERATION IS SICKLE CELL DISEASE

Same authors [2] defined SCD as hereditary hemolytic anemia resulting from the formation of abnormal hemoglobin (Hb S). Hb S differs structurally from the normal adult hemoglobin (Hb A) only in the substitution of valine for glutamic acid at the sixth position of the beta chain.

In the heterozygous state, i.e. sickle cell trait is due to the presence of HbS gene on one of the homologous pair of chromosome II, both adult and sickle hemoglobin (HbA and HbS) are present and the condition is usually asymptomatic except under extreme conditions like Hypothermia, Hyperthermia, Hypotension, Hypoxia, Hypo-volemia, Acidosis. This condition should be avoided .Which can leading to sequestration and thrombosis in the arterioles with subsequent ischemic infarction which may affect any organ in the body.

The same authors [3] consider sickle cell hemoglobinopathy a common genetic disorder affecting many individuals in certain countries. The disease is prevalent in the eastern and

south western provinces of Saudi Arabia. Saudi reports [4,5,6] found proportions of the various sickle genotypes in infants from Qatif and Al Hasa are very similar to comprehensive studies from Dammam and Qatif. Data from both studies indicate that in the Eastern Province, 20% to 30% of Saudi newborns are heterozygous for the sickle gene (FAS, FAS-Bart's), and 1.6% to 2.3% may be homozygous (FS, FS-Bart's). If, as is likely, about the same frequency of heterozygote is present in the adult Saudi population, it can be calculated that the frequency of homozygosis for sickle cell genes (SS) should be about one in 100 births, or 1.0 %

Electrophoresis study [7] reported on 2341 infants from the oases of Eastern Saudi Arabia, performed in an attempt to detect cases early and then to follow up and give better management to patients with sickle cell disease, showed 20% with S-trait and 43 infant with sickle cell disease (37 HbSS and 6 S-beta(0) thalassaemia).The incidence of Hb S for all Saudi Arabian neonates that were screened in one study[8] was 14.4% and ranged from 0.8% in Najran to 26.4% in AL-Qurayyat. HbS disease was present in 1.37% and sickle cell trait in 13.1% of all infants.

PERIOPERATIVE ANESTHESIA CONSIDERATIONS FOR SCD

Patients with SCD show wide variation in the severity from mild disease to very complicated with organ damage. The anesthesiologist should have full history and should search for evidence of end-organ damage in respiratory, cardiovascular, neurological, liver and renal. Investigations requested depends on the patients condition and if there is any symptom or sing of end-organ damage. The minimal of full blood count, electrolytes, renal function test, liver function test and chest x-ray. I.V. fluid should request during NPO period to avoid dehydration.

Hb should be > 7 mg/dl especially for the major surgery whine the bleeding is anticipated and to achieve Hb A ? 70% [9,10].

Inrtaoperative and postoperative, conditions can lead to acute sickling should be avoided dehydration, hypoxia, acidosis and low temperature.

THE SECOND CONSIDERATION IS MRI ENVIRONMENT

Definition and development of MRI was reported as following [11]

MRI is an imaging technique used primarily in medical settings to produce high quality images of the inside of the human body. MRI is based on the principles of nuclear magnetic resonance (NMR), a spectroscopic technique used to obtain microscopic chemical and physical information about molecules. The technique was called magnetic resonance imaging rather than nuclear magnetic resonance imaging (NMRI) because of the negative connotations associated with the word nuclear in the late 1970's. MRI started out as a tomographic imaging technique, that is it produced an image of the NMR signal in a thin slice through the human body. MRI has advanced beyond a tomographic imaging technique to a volume imaging technique.

Anesthesia considerations for MRI are: MRI compatible instruments. Relatively it is a long procedure. Cold environment is necessary to prevent heating the \MRI machine.. It is difficult to manage air way during procedure since the patient goes into the MRI tube and not easy to get access to the patient inside this tube. Claustrophobia and loud noise (up to 100 decibels) necessitate general anesthesia to children and properly maintain the airway. In our case we were able to help the radiology department to perform the procedure nicely without undue risk to the patient and allow the surgeon to have wise surgical decision. In this case was medical treatment to osteomyelitis of the spine beyond surgical intervention.

CONCLUSION

1. Sickle cell disease case can be done safely using hydration and worming accessible in BrainSuite.
2. BrainSuite which is next to the main Operating Rooms get the anesthesiologist more confidant due to facilities in the OR (Bair Hugger, holding area, Hot line-IV fluid warmer)
3. These cases need extra caution during anesthesia and extra time.
4. Good communication and good understanding between anesthesiologist, Anesthesia technicians, OR nurses and MRI technician, help to smooth safe procedure

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