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ANAESTHETIC MANAGEMENT OF A CHILD WITH OTAHARA SYNDROME PLANNED FOR LAPAROSCOPIC FUNDOPLICATION, FEEDING GASTROSTOMY AND UMBILICAL HERNIA REPAIR -A CASE REPORT.

GENERAL ANAESTHESIA IN AN OTAHARA SYNDROME CHILD - A CASE REPORT:

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Abstract:

Anaesthetic management of a child with Otahara syndrome planned for laparoscopic fundoplication, feeding gastrostomy and umbilical hernia repair.

Otahara syndrome is an uncommon, infantile epileptic encephalopathy with intractable seizures, refractory to anticonvulsants seen in the early neonatal period. It is diagnosed by the specific burst suppression pattern in EEG. Here we managed a 6-year-old child with Otahara syndrome for a large umbilical hernia repair. Meticulous precautions were taken to avoid all triggers,to maintain a seizure-free peri-operative period.

Keywords: Otahara syndrome, intractable seizures, general anaesthesia, umbilical hernia repair, lap fundoplication.

Introduction:

Otahara syndrome is an uncommon, epileptic encephalopathy characterised by abnormal EEG with intractable seizures in early infancy within the first 3 Months of age and often within the first two weeks.[1]It is synonymous with early infantile epileptic encephalopathy.[1] Infants develop focal or generalised frequent tonic spasms, either singly or in clusters, and are independent of sleep cycles. Spasms typically last for 10 seconds and can occur 10 to 100 times per day. It usually results in delayed psychomotor development and intellectual disability, associated with feeding difficulty, frequent respiratory and chest infections.[2]Although the birth history and head size are normal, microcephaly may occur. Aetiology lies in structural malformations, metabolic disorders, and genetic mutations.[2] Genetic variants manifest with additional signs such as tonic seizures, muscle stiffness, dilated pupils, ataxia, tetraplegia, and dystonia. Clinical presentation and typical interictal EEG show a burst suppression pattern comprising a burst of high voltage paroxysmal discharge separated by prolonged periods of nearly flat tracing that last for up to 18 seconds, which explains the diagnosis. Treatment consists of anticonvulsants and glucocorticoids to control seizures. Epilepsy surgery and

functional hemispherectomy contribute to surgical management. Prognosis is generally poor and the survivors manifest severe psychomotor impairment.[3]

Case Presentation :

A 6-year-old female child with Otahara syndrome who had a gross developmental delay and no gross and fine motor milestones with a GCS score of E4 V1 M5 presented with a large umbilical hernia. She was planned for lap fundoplication, feeding gastrostomy, and umbilical hernia repair. She was fed via NG tube, since 18 months of age and had multiple episodes of aspiration pneumonia and bronchopneumonia. She had intractable seizures of various semiologies which were noted and treated from the thirty second day of life. Seizures differed in character from early neonatal days to infancy and childhood such as quivering of lips, tonic posturing of upper and lower limbs, tonic jerking, vacant staring, and uprolling of eyes. Seizure attacks were noted even to the sound stimulus of switching on the light. Seizures started with repetitive clustered tonic spasms about 14 -15 times initially, later reduced to 3-4 times per day. Each episode lasted for about 20 seconds and resulted in the hypotonia of limbs with a loud cry or laughter at the end of an attack. She was on multiple antiepileptic drugs of various classes, including Levipill, Phenobarbitone, Clonazepam, Trihexyphenidyl, Parampanel, and Valproate, since her neonatal days.

On Examination

The child weighed 11 kg. She appeared dull, not responding to verbal and painful stimuli. She was microcephalic with isotropic- upward gaze, spastic limbs, b/l extensor plantar, and exaggerated deep tendon reflexes. She had less perception of pain. MRI showed mild cortical atrophy and EEG showed an abnormal pattern suggestive of epileptiform disorder with a burst suppression pattern in the occipital lobes.Her EEG showed sudden tonic posturing of the whole body lasting 2-5 seconds with eye deviation to either side with an electrical correlate of diffuse burst of spike and large amplitude slow wave followed by a diffuse attenuation lasting 8-10 seconds. Her neurosonogram, ABG and TMS were normal. DNA testing and genetic-chromosomal studies relieved heterozygous mutation variance of uncertain significance. Sequencing of variance in parents confirmed the mutation in genes.

On the preoperative day, she was strictly advised to continue all the antiepileptic drugs and to follow the NPO guidelines. On the morning of surgery,usual pre-medications were not given. Prilocaine 2% patch was applied 1h before on the left forearm. The plan of anaesthesia was general anaesthesia with controlled ventilation. A high-risk consent with an explained risk of peri-operative seizures with a postoperative ventilator consent was taken before the procedure. Heart rate, ECG, NIBP, ETCO2, and Spo2 were monitored. Meticulous precautions were taken to lower the pain stimulus. An IV access was secured over the LA prepared area. All the monitors were attached and baseline values were noted at the start of the surgery. A preheated warmer bed was kept ready to avoid hypothermia. All the pre-medication injections, Midazolam, Atropine, Fentanyl, and Paracetamol were given according to the weight. The child was started on, an injection of Levipill 500mg in 100 ml NS infusion, from the beginning of surgery. She was induced with thiopentone sodium and atracurium. The airway was secured with COETT of size 4.5 mm ID, and confirmed air entry.NG tube was inserted and confirmed. Maintenance of anaesthesia carried with propofol infusion at a dose of 8 ml/h and atracurium 0.5 mg/ml infusion at a dose of 4 ml/h throughout the surgery. Air and O2 were kept on flow. The intra-operative period was uneventful. We maintained a heart rate of 100-110 min, SPO2 100%, ETCO2 25-30, and respiratory rate of 18-20/min throughout the procedure. Local wound infiltration of inj.Bupivacaine 0.25% 10 ml was given to manage post-operative pain. After thorough suction, the child was reversed with neostigmine and atropine according to weight. She was extubated and shifted to Pacu for further monitoring.

Discussions:

There were multiple challenges, in giving general anaesthesia to this kid:

Here we managed a syndromic child with a disability to communicate and cooperate. Every precaution was taken to avoid pain and hypothermia which trigger seizure generation. All the painful stimuli were cautiously avoided. Drugs that increase seizure threshold potential were used for induction and maintenance of anaesthesia. Possible drug interactions due to long-term six anticonvulsants and their potential effects were dealt with cautiously. All the anti-epileptic drugs were continued along with the normal drugs until surgery. She exhibited resistance to neuromuscular blocking drugs as she was on long-term antiepileptic drugs. Propofol infusion and atracurium infusion were kept throughout the procedure.Inhalational anaesthetics and ketamine which decreases seizure threshold were avoided. Post-surgery, she was propped up and was

maintained on continuous NG aspiration to avoid regurgitation. Finally, all precautions were made to keep, a pain-free intraoperative and postoperative period to avoid central excitation on emergence from anesthesia.

Conclusion:

Otahara syndrome is a rare epileptic disorder with a low threshold of seizures.General anaesthesia in such patients needs great care in pre-operative planning, intra-operative pain, and anaesthetic management as well as meticulous use of anaesthetic drugs.

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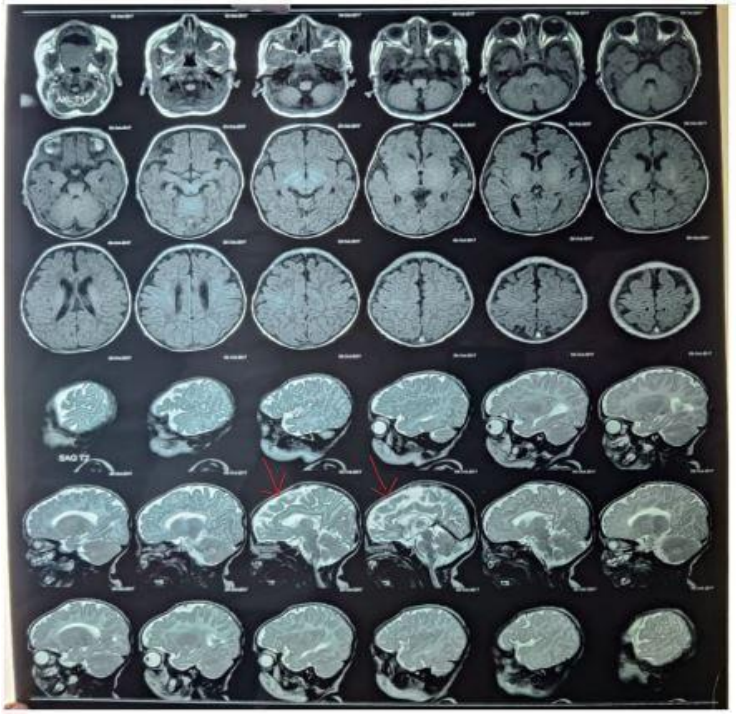

Figure 1: Mid-section MRI with diffuse brain atrophy shows increased sulcal space and decreased brain parenchyma.

Declaration of patient's consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given consent for her images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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