CASE REPORT

Anaesthetic Challenges in Neuronal Ceroid lipofuscinoses for Percutaneous Endoscopic Gastrostomy

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Website:
theanaesthesiologist.com

Authors:

Dr Nisha Rajmohan, Senior Consultant Anaesthesiologist, Department of Anaesthesia and Critical Care, Astermedcity ,Kochi.
ORCID 0000-0003-1389-179X

Dr. Abin K Jose, Fellow in Cardiothoracic Anaesthesia Department of Anaesthesia and Critical care, Astermedcity, Kochi, Orcid id 0009-0005-1497-3414

Dr Balu Sankar J, Specialist, Department of anesthesia and Critical care Astermedicity ,Kochi.
ORCID 0000-0003-2465-7456

Dr Sunitha Chidambaran Thankam, DA, Specialist, Department of anesthesia and Critical care, Astermedcity ,Kochi.
Orcid id 0009-0003-5534-3430

Dr Suresh Gangadharan Nair, Lead Consultant, Department of Anaesthesiology and Critical care, Aster Medcity, Kochi.
ORCID 0000-0003-3633-4179

Corresponding Author:
Dr Nisha Rajmohan,
Senior Consultant Anaesthetist, Astermedcity,Kuttisahib Road, Cheranalloor, South Chitoor, Kochi,Kerala - 682027, India.Phone Number: +919656732203 Email: nishavismaya@gmail.com

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Neuronal ceroid lipofuscinoses are rare inherited lysosomal storage diseases with multisystem involvement. Here we describe the anesthetic management of a six-year-old female patient who presented with recurrent aspiration pneumonia, multidrug resistant seizures, progressive deterioration of milestones. She was mute with impaired vision and was posted for Percutaneous Endoscopic Gastrostomy. Presence of uncontrolled seizures, difficulty in communication, cognitive impairment, predisposition to bradycardia, hypothermia, risk of aspiration with persistent lung infection were few of the challenges we faced. Optimization of seizure therapy, antibiotics, chest physiotherapy, nebulization with broncho dilators helped in preoperative preparation. Use of glycopyrrolate helped prevent bradycardia and reduce secretions. Propofol and midazolam for induction of anaesthesia helped prevent seizures during the procedure which was done under intravenous sedation with monitored anaesthesia care. Patient was positioned slight head up to prevent aspiration and we were ready to intubate if necessary. Warmers and warm fluids helped prevent hypothermia. The procedure was completed uneventfully.

Introduction

Neuronal ceroid lipofuscinoses (NCL) are a group of autosomal recessive inherited neurodegenerative diseases characterized by accumulation of lipofuscin in tissues. [1] Clinical manifestations are progressive mental and motor deterioration, multidrug resistant seizures, visual loss, cardiac involvement with intra operative arrhythmias (bradycardias) and hypothermia.[2] We report the anaesthetic

challenges in managing a six-year-old female child with NCL for percutaneous endoscopic gastrostomy (PEG).

Case report

Six-year-old female child, (weight 18.7kg) presented with recurrent aspiration and pneumonia. She was diagnosed as a case of NCL 7 at 5 years by genetic study. She was the second child of 3rd degree consanguineous parentage was delivered by LSCS with a birthweight of 2.4kg. The child was apparently normal till 3 years of age when she developed abnormal movements in the form of drop attacks, progressive deterioration of all her attained mile stones and recurrent aspiration pneumonia. From the past one year she was bed ridden, mute with impaired vision with no fixing or following. She was admitted to our hospital following aspiration pneumonia with desaturation (SpO2 85%) and was started on high flow nasal canula (HFNC). Chest x ray showed multiple opacities on right side suggestive of aspiration. She had pooling of secretions following feeds and required intermittent suctioning. Pediatric neurology consultation was done in view increased dystonic movements. Her clobazam dose was increased from 0.5mg/kg/day to 1mg/kg/day and Sodium Valproate was continued at 20 mg/kg/day. The child was started on nebulizations with salbutamol sulphate and ipratropium bromide and budesonide, intermittent suctioning, physiotherapy and antibiotics. She was weaned off HFNC to O 2 via pediatric face mask at 5l/min. multidisciplinary team meeting was conducted and the parents' concerns were addressed. They were advised trial of percutaneous endoscopic gastrostomy (PEG) as a supportive measure for feeding. The plan was PEG under monitored anesthesia care with intravenous sedation after taking a high-risk consent. All the regular medications were continued in the morning. She had crepitations, more on the right side. O2 2-3l/min was administered with nasal prongs; patient was positioned supine with head turned to the left side and slight reverse Trendelenburg position 15 degree was given. Electrocardiogram, SpO2, noninvasive blood pressure, temperature monitoring was done. Patient was given glycol -pyrrolate 0.1mg, fentanyl 20mcg, midazolam 0.25mg and propofol 10mg. We had to repeat dose of propofol 5mg twice to maintain the depth of anaesthesia, PEG tube insertion was done after infiltration of puncture site with 5ml bupivacaine 0.2% infiltration. Warmed plasmalyte with 2% dextrose was given at the rate of 60 ml/hr. Hot air warming blankets helped in maintaining the temperature. After the procedure was completed uneventfully, she was shifted to pediatric intensive care unit for monitoring. The mother was taught how to suction, give PEG feeds and do physiotherapy The child was discharged on the 7th post procedure day.

Discussion

NCL is associated with progressive multiorgan dysfunction. Loss of airway tone with drooling of saliva, lower oesophageal sphincter dysfunction, repeated aspiration pneumonia, ongoing dystonia, chance of prolonged ventilation and failure to wean called for proper planning for anesthesia. [1]
Adequate control of respiratory infection and dystonia was one of the first steps towards this goal. Lack of communication with poor muscle tone rendered incentive spirometry impossible. [1,2]

Even though this patient had a risk of aspiration, we avoided intubation as there is a possibility of prolonged ventilation with attended problems. It was a short procedure which normally does not call for endotracheal intubation. [3,4] A slight head up position of 15 degree was used to avoid regurgitation. We were ready to intubate the patient in the event of respiratory compromise. Glycopyrrolate reduced secretion. Use of intermittent propofol also helped in maintain the depth of anesthesia at the time of introduction of endoscope and helped in the early recovery of this patient with hypotonia.[2] Propofol and midazolam with its antiepileptic action helped in preventing triggering a seizure. The use of local anesthesia at the site of incision helped achieve adequate analgesia.[1]

Since they are prone to hypothermia, proper warming blankets, warm fluids helped maintain

temperature for this very short procedure(20min). [2] Bradycardia was avoided by avoiding drugs causing bradycardia like dexmedetomidine and also by giving glycopyrrolate.

CONCLUSION

NCL due to multisystem involvement and rarity present a challenge to the anesthesiologist. Meticulous preoperative preparation, awareness of anticipated complications can help in proper management.

Keywords: Neuronal ceroid lipofuscinoses disease, Gastrostomy, Children, Endoscopy

REFERENCES

1. Kako H, Martin DP, Tobias JD. Perioperative care of a patient with neuronal ceroid lipofuscinoses. Saudi Journal of Anaesthesia. 2013; 7:336.
2. Yamada Y, Doi K, Sakura S, Saito Y. Anesthetic management for a patient with Jansky-Bielschowsky disease. Canadian Journal of Anesthesia. 2002; 49:81.
3. Luckett A, Yousef M, Tifft C, Jenkins K, Smith A, Munoz A, Quimby R, Porter FD, Dang Do AN. Anesthesia outcomes in lysosomal disorders: CLN3 and GM1 gangliosidosis. Am J Med Genet A. 2023; 191:711-17.
4. Parker JD. Pulmonary aspiration during procedural sedation for colonoscopy