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Case report

Anaesthetic Management of a sickle cell trait child with congenital insensitivity to pain and Anhidrosis: A case report.

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Abstract:

Congenital insensitivity to pain with anhidrosis(CIPA) is extremely rarae autosomal recessive hereditary sensory neuropathy which affects pain sensation, thermoregulation and present with various degree of mental retardation.Due to its rare presentation anaesthetic management of such patients pose a challenge for Anaesthesiologists. In India very few cases have been reported till date. We are hereby reporting a case of 9 year old girl having sickle cell trait with CIPA posted for sequestrectomy of right Ankle.

Introduction

CIPA syndrome was first described by Nishida in 1951, an autosomal recessive Hereditary autonomic and sensory neuropathy type IV also known as Nishida syndrome".[1,7] The clinical presentation comprise of absence of sweating, insensitivity to pain and touch, recurrent episodes of fever, self mutilating behaviour in addition to mental Retardation are pathognomonic features.[1,2,8] Varying degrees of sensory loss including nociceptive hyposensitivity and various degrees of autonomic dysfunction is a common presentation . Chronic osteomyelitis is one of the possible manifestations of CIP, which can be difficult to diagnose and treat due to the lack of pain as a diagnostic criterion.[9]

Case Description:

A 9 year old girl weighing 16kg presented for pre anaesthesia check up with chief complaints of pus discharge from right ankle since 18 months followed by an injury she was unaware of. She was diagnosed as chronic oseteomyelitis of ankle bone Further history revealed that she has no pain response and having self mutilation injuries are present on the nose, lips and fingers , she also has widespread fissure on skin and fingers due to dryness and absence of sweating. (figure 1,2) Her family history was insignificant being born full term without any mental retardation which was consistently seen with this hereditary sensory and autonomic neuropathy IV and was 3rd child of the non-Consanguineous

parentage whereas other siblings were normal. History of multiple blood transfusions reported in past Hb electrophoresis revealed sickle cell trait.Past medical history also revealed frequent episodes of fever. Despite her insensitivity to pain when assessed she flinched on deep noxious stimulus. The child was diagnosed with CIPA based solely on clinical symptoms. 2D ECHO was done to rule out cardiovascular abnormality.

ANAESTHETIC MANAGEMENT:

Written and informed consent were taken from parents explaining the risks involved . All blood investigations were well within normal limits. Child was kept NPO for 6 hrs, , and overnight hydration was maintained with iv fluids. On arrival to the operation room standard ASA monitors were attached and baseline Heart rate, NIBP, Oxygen saturation , Body temperature and ECG is noted. Operation room temperature was kept between 23-25 degrees, IV fluids were cooled beforehand. Patient was premedicated with injection Ondansetron 1.5mg, injection Midazolam 0.5 mg, and injection Glycopyrrolate , as it was a short procedure Total intravenous Anaesthesia was planned with injection propofol 1-1.5 mg/kg given as bolus first and anaesthesia was maintained with 25-50 micrograms per kg minute infusion via infusion pump titrated to keep Mean arterial pressure up to 65 mm hg. Patient was spontaneously breathing 100% oxygen given through Jackson-Rees circuit . we avoided opioids and inhalational Agents as it was a short procedure to ensure early recovery and also to minimize post operative complications . Injection paracetamol 240 mg given in infusion for analgesia. Intraoperative vitals , temperature and end tidal co2 was monitored and no major fluctuations noted. Body temperature was between 36-37 degrees during procedure and no extra measure done to control body temperature. The procedure lasted for 30 mins. No unpredictable events like hypotension, bradycardia or hyperthermia noted in our case. Her recovery was smooth , after 5-10 mins she was responding well to the commands .she was shifted to Post operative care room for monitoring.

Discussion

The mutation in NTRK1 hinders the ability of nerve growth factor to bind to the receptor properly, leading to the characteristic lack of nociceptive reception.[4] Consequently, patients with CIPA are susceptible to numerous anaesthetic complications due to autonomic abnormalities, including hemodynamic instability, particularly hypotension, bradycardia, and hyperthermia[5] Due to the rarity of CIPA, anesthesia management experience is limited, Cases of CIPA often present to the hospital with orthopedic symptoms from repeated trauma resulting in Charcot joints and osteomyelitis, at times requiring surgical intervention.[6] Half of the reported cases of CIPA were seen in consanguineous parentage and some degree of mental retardation was also seen which makes our case unique as our patient was born to a non consanguineous couple without any mental retardation .[4]

She was very cheery and happy child without any developmental delay. To tackle unpredictable events that can occur in this case which is still a challenge for anaesthesiologist. our anaesthetic management focused on three things a)To maintain normothermia b). To minimize anaesthetic drug dosage & exposure 3). To maintain cardiovascular and hemodynamic stability.[1]

Majority of studies have given regional as well as general anaesthesia. successfully. We have given midazolam for anxiolysis. Some of the CIPA patients reported hyperthermia[2] but our patient was euthermic throughout the procedure . As our patient was also a sickle cell trait we maintained her hydration and oxygenation preoperatively as well.

We have not used any opiates in our study which is different from other studies where they used fentanyl to blunt the response during intubation , our patient was not intubated so we avoided opiates and volatile agents. We have not seen any bradycardia or hypotension in our study but few of the researchers have found bradycardia with profound hypotension in their study[5]**.** Majority of cases have induced anaesthesia with propofol along with muscle relaxants but due to shorter duration of surgery we have chosen TIVA for early awakening and recovery. For analgesia we have used injection Paracetamol . we did not have any intraoperative and postoperative complications . the child recovered quickly from anaesthesia and shifted to PACU for further monitoring. No event of post operative nausea and vomiting noted.

CIPA syndrome requires multidisciplinary approach and educating the parents about the complications associated with it.

CONCLUSION : We hereby conclude that Anaesthetic implication in CIPA cases requires keen vigilance and awareness of the anticipated complications associated with autonomic dystonia and temperature dysregulation and airway management. Pain insensitivity can be of varying degree which requires proper depth of anaesthesia and analgesia intraoperatively as well as in Post operative period.

Conflict of interest : none

Consent: consent was obtained from parents.

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Figure 1. joint deformity of hand and self mutilations.

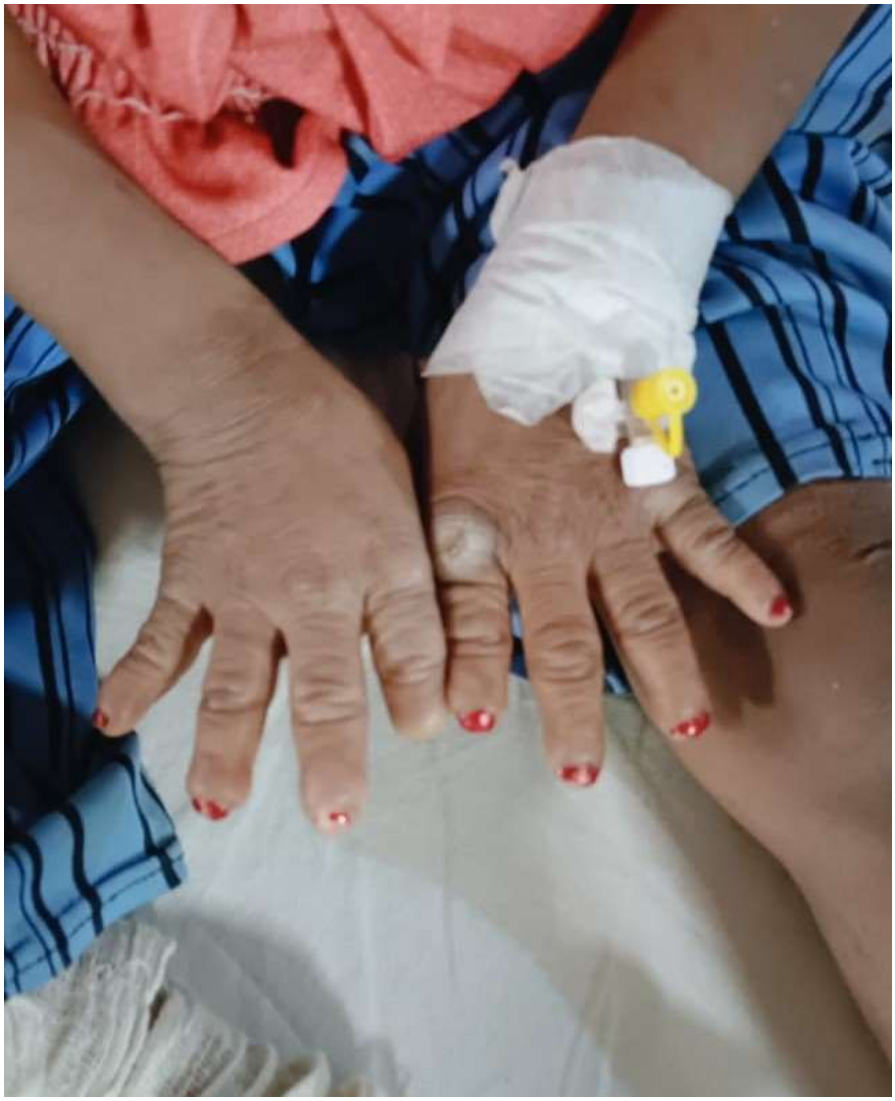

Figure 2. selfmutilations of nose, dry skin , fissures due to lack of sweating