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Case Report

Expect the Unexpected - Peri-Operative Challenges for the Anesthesia Team During Management of Kenny- Caffey Syndrome - A Case Report

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Abstract

Kenny - Caffey syndrome is a extremely rare syndrome . The total number of documented cases in the world are less than 100 patients with 60 patients living in the MENA region (Middle East North Africa) & the rest related to Middle eastern origin. These patients present unique challenges to the medical profession including altered growth patterns with fragile body habitus , endocrine disorders , severe hypocalcemia & mental retardation.Regular outpatient visit reasons include micrognathia with dental caries as is microopthalmia with hypermetropia. We present a case report & the challenges faced by the Anesthesia team in a 21 year female patient coming for urgent Laparotomy with a diagnosis of bowel obstruction(sigmoid volvulus)

Keywords: HME- humidifier & moisture exchanger, OPD- outpatient department

PE - phenyl ephrine , MAC - minimum alveolar concentration, PRBC- packed red blood cells , FFP - fresh frozen plasma, MDT- muti disciplinary team.

Introduction

The condition was originally described by Frederic Kenny and Louis Linarelli in 1966 . In 1967, John Caffey, a pediatric radiologist, described the detailed radio-graphic findings of various bones in the same family. The condition was called the Kenny-Caffey Syndrome. A somewhat similar, but recessively inherited condition known as Kenny-Caffey Syndrome Type 1 (KCS1) also exists. This recessive condition includes intellectual disability as a common symptom, and is also known as the Sanjad-Sakati syndrome or Hypoparathyroidism-Retardation-Dysmorphism Syndrome which is the common variant in the MENA region. The total number of patients in the UAE is unknown there are occasional inter hospital reports of KCS(Type 1) patient visits mostly from the same family in our Health system database.

The Case Report

A 21 year female patient was posted for urgent laparotomy with a possible colostomy . A diagnosis of large bowel obstruction as per the medical history, clinical signs & symptoms and a CT scan. The electronic records of the hospital showed this a first visit to our tertiary care centre & the recorded weight of the patient was 10.5 kgs. Since the patient had mental retardation all history was obtained from the parents of the patient.

On examination: - the patient presented with microcephaly & webbed neck with irregular dentition, small body habitus & deranged lab values & the records from the previous hospital showed regular opd visits for dental caries. There was no previous history of any surgery or any anesthesia record for any diagnostic procedure. Laboratory investigations showed moderate anemia 8 gm/dl & normal platelet count with severe hypocalcemia 2.6 mg/dl& serum potassium values of 2.95 mmol/l. The ECG showed severe tachycardia & recorded vitals showed BP 105/75 mm HG with HR of 140 bpm. Due to pain & mental retardation the patient was restrained in the ward. A quick auscultation of the heart showed regular heart sounds & no cardiac murmur was detected. Airway was presumed as Mallampatti 4 in view of microcephaly & lack of cooperation from the patient. Medical treatment from childhood included Parathormone injections. Vitamin D3 & calcium supplementation.Since it was a rare case an urgent multidisciplinary treatment approach was planned via video conference & all stake holders involved (surgical team - anesthesia team & nursing teams from both the Operating room and surgical icu) had a quick discussion about possible outcomes & a plan was made regarding the Peri-operative journey of the patient in our Hospital.

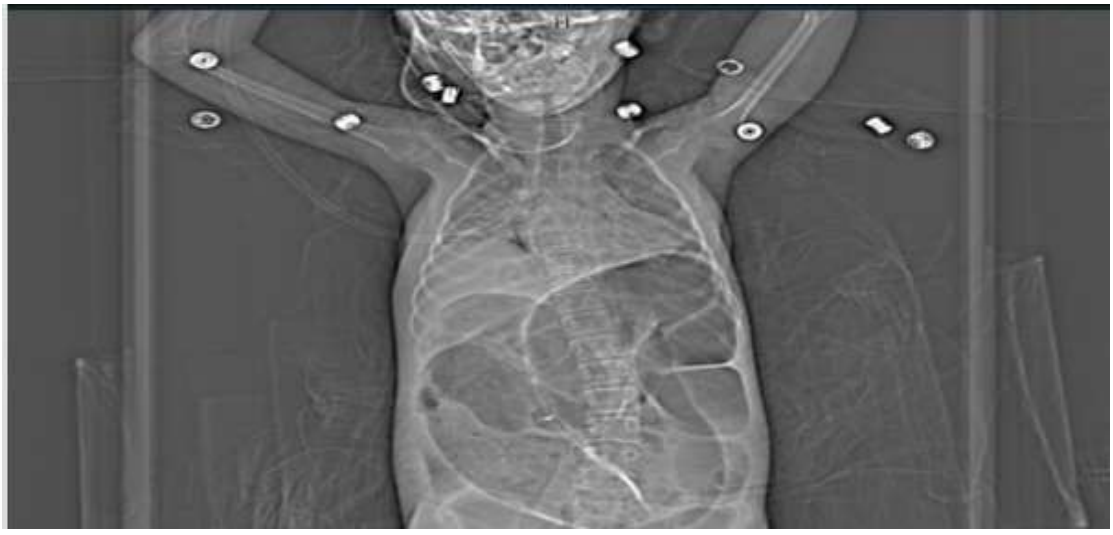


Image 1 shows shows preoperative CT scan

Findings: Significant colonic distension with faeces and fluid in the large bowel with the marked distension of the sigmoid colon with an apparent kink of the distal sigmoid colon and even though it is an unusual pattern, appearances favour a distal sigmoid volvulus.

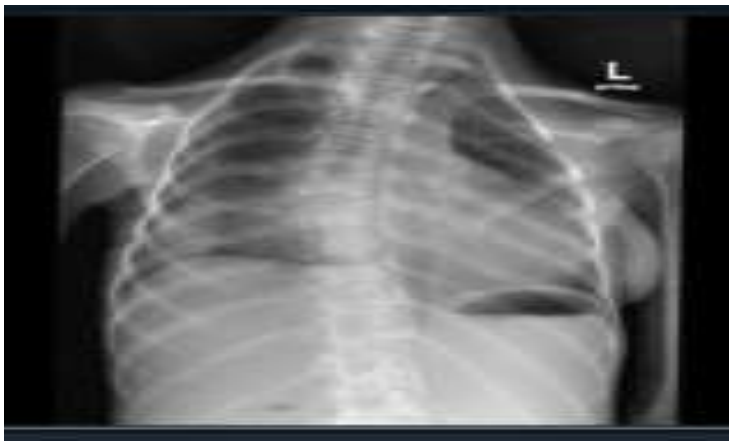


Image 2 --Chest X ray findings -ground glass consolidation in the lungs.

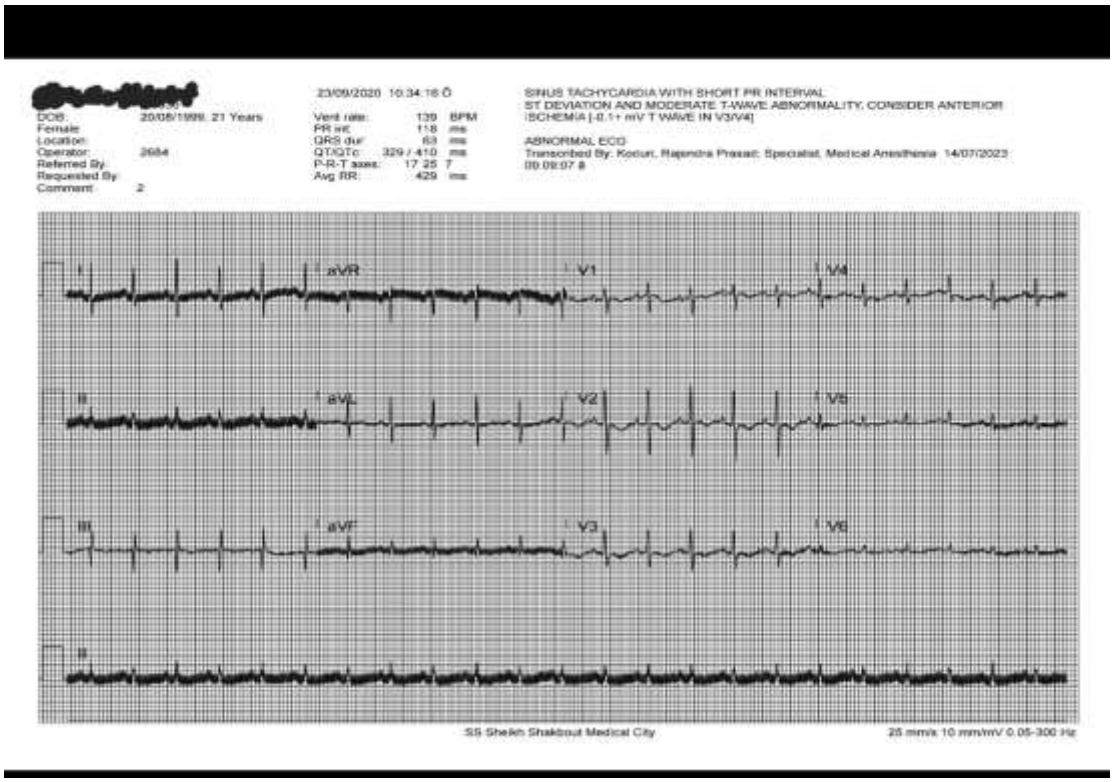


Image 3 - Preoperative ECG -- shows sinus tachycardia

Challenges faced by Anesthesia team during the Peri-operative period.

This 21 year patient (adult by age) is only 10.5 kgs (pediatric <20 kgs) with extreme hypocalcemia.

1- Urgent correction of serum calcium from the existing 2.6 mg/dl to 7 mg/dl.

2- Change of adult operating table to pediatric table

3- Change of adult circuits to pediatric circuits

4- Change of adult HME to pediatric HME on the anesthesia circuits

5- Change of ventilator status from adult to pediatric mode

6- Under body warmers as in the pediatric patient (although the patient by age is an adult) to prevent hypothermia & increase in ambient temp of the o.t.

7- Airway challenges --proper sized mask & keep the difficult airway trolley ready including video-laryngoscopes.

8- Restrain the patient as in the ward & adequate padding of all pressure points to prevent iatrogenic fractures during induction.

9- Proper weight settings on the syringe pumps for the ionotropes.

10- Setting up Ultrasound in the room for introduction of invasive lines (arterial / cvp line) for monitoring of acid base balance -& monitoring serum calcium.

11- Surgical team should include a ENT consultant for emergency tracheostomy if necessary & a pediatric surgeon to handle the undersized organs.

12- Resuscitation of patient using 5% albumin rather than crystalloid to maintain vitals within normal limits.

Peri-operative course:

Surgical team had estimated the course of surgery for 2-3 hours which included Lapartotomy with possible colostomy. The patient was received in the holding bay & received 150 mg iv calcium gluconate infusion in 30 minutes

Serum calcium improved to 7 mmol with calcium gluconate infusions under ECG monitoring. The preoperative S. Potassium was 2.97 & the inverse relationship of potassium & calcium levels was considered

before more calcium gluconate infusion. Since the patient was used to lower levels of Serum calcium we used the lower end o f the normal range for correction. Serum albumin levels were noted at 2.9 g /dl. Adequate hydration with dextrose saline @ 50 ml/hr was instituted in the ward. Necessary blood & blood products were crossmatched for the surgery.

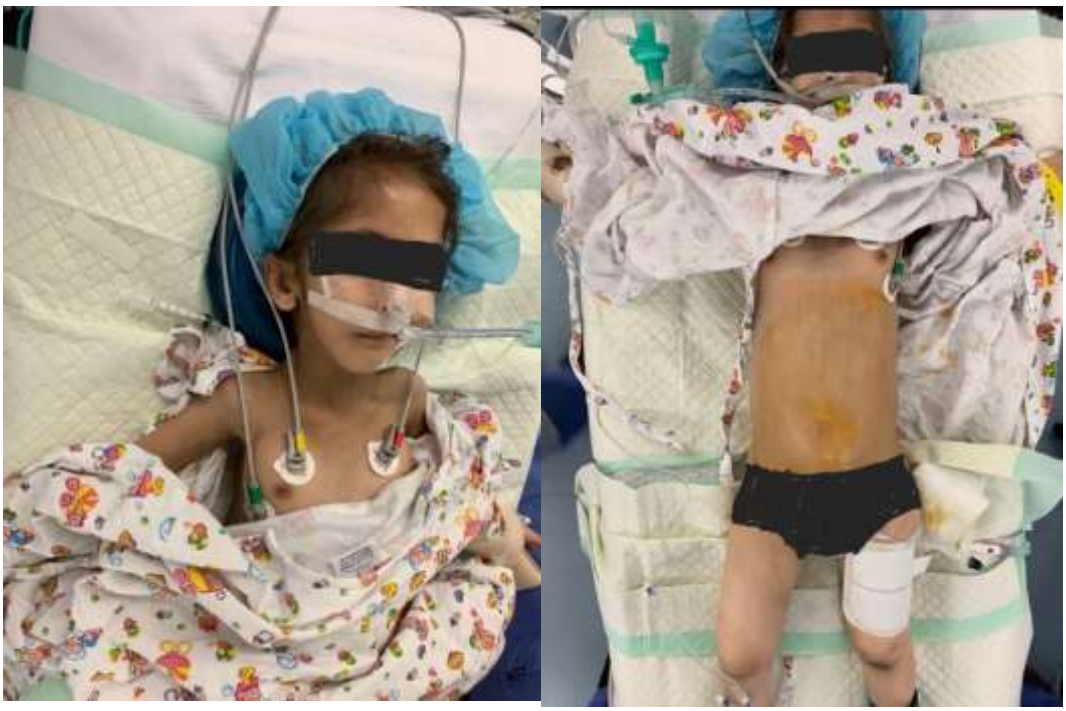
In the ot Ketamine 10 mg iv was given to facilitate attachment of regular monitors to this uncoperative patient. (EKG, NIBP & Pulsoximeter)

Standard induction with rapid sequence intubation using propofol 2mg/kg fentanyl 1 mic/kg & rocuronium 1mg/kg was planned Cricoid pressure was used to prevent regurgitation of stomach contents in this case of intestinal obstruction. Videolaryngoscopes were difficult to introduce because of adult dentition in a micrognathia situation. Classic Mc coy laryngoscopy with size 1 blade was used & size 5 cuffed oral endotracheal tube was introduced with difficulty (irregular dentition & micrognathia) by the 2 nd attempt. A 8 FG nasogastric tube was introduced after induction& all gastric contents were aspirated.

During induction 10 micrograms of phenylephrine were given prophylactically to prevent induction induced hypotension. Maintenance drugs of fentanyl & relaxant were weight based in this case.

Positioning for invasive lines in this fragile patient was very difficult & ultrasound guided introduction of invasive lines included right subclavian line & left arterial line for Peri-operative monitoring of acid base balance & serum electrolytes. Induction induced hypotension was treated with PE infusions. The patient was ventilated with 40 % oxygen / Air with sevoflurane titrated to 1.2 MAC. Vitals were maintained through out with necessary colloid infusions 5% albumin & PE infusions. Hypokalemia was corrected to 3.5 mmol to prevent dangerous arrhythmia. Serum glucose & body temperature were maintained at normal levels. One unit of warmed PRBC & 1 unit of FFP was transfused during the Peri-operative period.

Surgeons were gently reminded about fragility of the patient & gentle movements during surgical retraction of abdomen was done. Surgery (laparotomy with ileocolic anastomosis) proceeded uneventfully the patient was shifted to the SICU for post operative elective ventilation .



Post op course: In the SICU elective ventilation was commenced as per MDT plan for the first operative day with maintainence of vitals using nor epinephrine infusions (dose as necessary) & sedo-anlagesia using fentanyl/ Ketamine infusions. The patient was weaned away from the ventilator by 24 hrs of admission & extubated . Monitoring in the SICU proceeded along with regular lab investigations & the patient was shifted to the ward on the 4 th post operative day.

Conclusion

Kenny Caffey is a very rare syndrome but not exclusive to the MENA region. Anesthesia teams may face this patients once in a life time .Apart from opd visits we do not have any published literature regarding the Peri-operative challenges faced by the anesthesiologists & this is the first such instance. Proper preoperative evaluation of the patient with focus on correcting deranged lab values, liver function tests (especially serum albumin) &quick securing of airway will prevent crisis situations. Proper planning & discussion with MDT approach is so important for successful outcome.

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