Pediatric Anesthesia and Critical Care Journal 2014; 2(1):52-54
doi:10.14587/paccj.2014.13

PACCJ

Diffuse cerebral atrophy, central sinus venous thrombosis, hypoplastic carpus callosum, ADHD and VDS in a child: anaesthetic implications

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Key points

Attention deficit hyperkinetic disorder (ADHD) is characterized by inattention, poor impulse control, motor overactivity and restlessness.The clinical features most commonly seen in patients with hypoplastic carpus callosum syndrome are microcephaly, seizures, developmental delay or retardation, hypotonia, spasticity and multiple somatic anomalies including arthrogryposis, eye anomalies, syringomyelia in the spinal cord and facial dysmorphisms. The response to general anaesthesia with sevoflurane in this rare complex combination of disorders and its postoperative outcomes were observed.

Abstract

Central venous sinus thrombosis (CSVT) in neonates, infants and children is often multifactorial in etiology, with a predisposing comorbid condition or infirmity identified in up to 95% of those affected[1]. These conditions include common childhood illnesses such as fever, infection, dehydration, and anemia, as well as acute and chronic medical conditions such as congenital heart disease, nephrotic syndrome, systemic lupus erythematosus and malignancy[1]. Attention deficit hyperkinetic disorder(ADHD) is the most common neurobehavioral disorder of childhood with an incidence of 5% in school children. It is characterized by inattention, poor impulse control, motor overactivity and restlessness[2]. We report a case of a 2.5 year old female child with diffuse cerebral atrophy, hypoplastic carpus callosum with central sinus venous thrombosis, ADHD and ventricular septal defect posted for sqint surgery of both eyes under general anaesthesia with sevoflurane.

Keywords: ADHD, central sinus venous thrombosis, sevoflurane, pediatric.

Case report

A 2.5 year old female child weighing 10 kg posted for squint surgery of both eyes under general anaesthesia. She was born with full term caesarian delivery, cried immediately after birth weighing 2.8 kg. Child had microcephaly, subtle dysmorphic features, generalized hypotonia, poor feeding, global developmental delay and failure to thrive. At the age of 8 months, she was diagnosed with congenital heart disease having ventricular septal defect with severe pulmonary arterial hypertension, large post tricuspid shunt and cardiac failure. She was operated for the same 17 months ago. Child had complex postoperative course. Patient developed seizures and antibiotic induced enterocolitis due to sepsis that was treated conservatively. CT scan of brain showed diffuse cerebral atrophy, hypoplastic carpus callosum, acute on chronic central venous sinus thrombosis par-

tially recanalised and subdural hygroma in left frontoparietal region. Child was put on anti epileptic drugs and heparin followed by warfarin. There was difficulty in weaning in postoperative period. Patient was on ventilator for 40 day with tracheostomy and inotropic drugs and in intensive care for almost three months. Tracheostomy scar was closed, gradually condition improved and she was discharged after 98 days. At present, child was posted for squint surgery(four muscles) of both eyes under general anaesthesia. Blood investigation revealed normal hemogram, renal and liver profile. TORCH showed positive for Ig G antibodies of CMV and Ig Mantibodies of rubella suggesting congenital infection. Ultrasound abdomen was normal. CT Scan of brain showed hypoplastic carpus callosum, diffuse cerebral atrophy, resolving sub dural hygroma and chronic sinus venous thrombosis with partial recanalisation. ECHO shows small VSD with no pulmonary arterial hypertension and good biventricular function. She was not any cardiac drugs or warfarin. Child was undergoing behaveriol and medical (only nutritional) therapy for ADHD. She was seizure free from last 6 months. She was able to walk and speak. ENT opinion was sought and the trachea was clear and devoid of any adhesive bands. Patient was taken as first case with standard fasting protocols. Parent was accompanied with the patient in the operation theatre before induction. All parameters like Electrocardiography non invasive blood pressure, EtCO2 and temperature probe was applied. Inhalational Induction was done in distraction technique with sevoflurane in an incremental manner quickly upto 8 vol % via facemask.

Intravenous line was secured. Intravenous glycopyrrolate 0.06 mg/kg, fentanyl 15 mcg was given. After making sure of adequate ventilation with the bag and mask, atracurium 5 mg was given and ventilated with 100% oxygen. Direct laryngoscopy was done and trachea was intubated successfully in a second attempt. Patient was catheterised. All vital parameters were monitored throughout the surgery.

Anaesthesia was maintained with oxygen, nitrous oxide and sevoflurane. Procedure took three and half hours. Patient tolerated anaesthesia well. Child was extubated after the child was completely awake. Propofol one mg/kg was given ten minute before extubation to prevent emergence delirium. Intravenous methylprednisolone 10 mg and ondasetron 0.1 mg was given intravenously. Child was monitored for seizures, bradycardia and other hemodynamic parameters in a quiet intensive care environment for 24 hours. Child had two episodes of vomiting in the postoperative period and required sedation with midazolam 0.05mg/kg once in the night. Patient was discharged on third postoperative period.

Discussion

The incidence of childhood CSVT varies between 0.4 and 0.7 per 100,000 children per year[3] CSVT in infants and children is often multifactorial in etiology. These conditions include common childhood illnesses such as fever, infection, dehydration and anemia, as well as acute and chronic medical conditions such as congenital heart disease, nephrotic syndrome, systemic lupus erythematosus, and malignancy[1]. CSVT has also been reported in chronic anemias, such as hemolytic anemia and Evans syndrome[4]. Dehydration and hypovolemia should always be carefully assessed and corrected to prevent thrombus propagation and promote recanalization of the affected vessel [1]. The clinical manifestations of CSVT are nonspecific, may be subtle[1] and may overlap with predisposing conditions such as infection and dehydration.[5] Seizures, altered levels of consciousness and encephalopathy, focal neurologic deficits and diffuse neurologic symptoms (headache, nausea, emesis) may result. CSVT-specific mortality is less than 10%, but motor and cognitive sequelae may require long-term rehabilitative regimens[5]. Coma is a predictor of death in childhood CSVT.[5] Attention Deficit Hyperkinetic Disorder (ADHD) is the most common neurobehaviuoral disorder of childhood with an incidence of 5% in school Children characterized by inattention, poor impulse controlmotor overactivity and restlessness. Inadequate do-

pamine and noradrenaline in the fronto-subcortical-cerebellar regions may cause under stimulation of inhibitory pathways[2]. The effectiveness of stimulants in treatment supports this theory poses similar problems to anaesthetists as autistic spectrum of disorders. [2]

Perioperatively, children tolerate poorly waiting long periods in hospitals and behavior may become disruptive on the ward, minimize waiting times wherever possible, plan to do the case in the day when the child is more cooperative (ideally, first on a morning listproviding a quiet room to waiting can reduce preoperative anxiety and adverse behavior. Distraction techniques may help when the child is waiting and at induction of anaesthesia. Sedative premedication is used frequently in this group of patients and effect are less predictable and more variable in more variable in patients on stimulants. Medication modify noradrenergic and dopaminergic functions in CNS. They may reduce the seizure threshold and predispose to PONV. Stimulants like methylphenidate may increase the MAC value of anaesthetic agents.[2] Difficult inubation was anticipated. ENT opinion was sought to rule out any bands within the trachea because of tracheostomy. Fibre optic guided intubation can be very helpful. Use of depolarizing relaxant was restricted in view of malignant hyperthermia and hyperkalemia. Dehydration was be avoided in view of central venous sinus thrombosis. It was taken as first case and adequate clear liquids can be allowed two hours prior to surgery. Euvolemia was maintained peri and post operatively. Prophylactic phenytoin can be given to avoid seizures. Heart rate was monitored over 24 hours for bradycardia as both the medial rectus was operated. Prophylactic steroid was given to reduce postoperative oedema of operated extraocular muscles and bradycardia. Recovery should be smooth.so propofol one mg/kg ten minutes before extubation to reduce the emergence delirium. Child may require sedation in intensive care because of ADHD. It was carefully titrated according to the need.

Conclusions

Central venous sinus thrombosis and ADHD are major issues of anaesthetic concern in our case. Accompanying parents before induction, ruling out any adhesion band in trachea, anticipating difficult intubation, maintaining good hydration according to central venous pressure, reducing emergence delirium by giving propofol if sevoflurane used, monitoring heart rate in postoperative period, adequately preventing and treating PONV, maintaining a quite and friendly environment in the intensive care and sedating the child according ot the need will come a long way in successfull management of these type of cases.

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