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Case Study

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ANAESTHESIA IMPLICATIONS IN CORPUS CALLOSAL AGENESIS

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ABSTRACT

Agenesis of corpus callosum is characterized by partial or complete absence of interhemispheric fibres connecting the two cerebral hemispheres. It may be inherited as an autosomal recessive trait or an X linked dominant trait. This condition has varied symptoms such as epileptic seizures, feeding problems, delayed developmental milestones, visual and auditory impairment, posterior urethral valves, etc. Anaesthesia given to such patients has its own challenges. We present a case of corpus callosal agenesis with posterior urethral valves. Patient underwent initial fulguration and later a bilateral ureteric implantation under general anaesthesia. A brief case report and anesthetic implications are presented along with review of literature.

KEYWORDS: Anesthesia, corpus callosal agenesis, ureteral implantation.

INTRODUCTION

Corpus callosal agenesis is heterogenous in etiology. The incidence in autopsy series is reported to be 1 in 20,000.[1] Among pediatric patients referred for magnetic resonance imaging (MRI) because of neurologic abnormalities, the incidence is 1 in 100 or greater. The defect is more common in males than in females with a sex ratio approaching 2:1.

Its occurrence has been documented in the fetal alcohol syndrome[3] and other teratogenic causes such as maternal diabetes and infectious agents have been suggested in isolated case reports.[2,3] Inborn errors of metabolism are an important cause of agenesis of the corpus callosum and may represent up to 4% of all cases.[1]

Corpus callosum begins to form as early as 6 weeks of gestation. At 11-12 weeks of gestation, the first fibres cross the midline through the massa commissuralis, to form the corpus callosum. There are two primary types of agenesis. In the first type, the callosal axons develop and move toward the midline but do not cross. This results in the formation of the longitudinally oriented bundles of Probst that are located medial to the lateral ventricles in patients with this disorder and are pathognomonic of the defect. In the second type, the commissural axons or their cell bodies fail to form and approach the midline. This is considerably less common and usually associated with syndromic forms of agenesis of the corpus callosum.

These patients may present with associated congenital posterior urethral valves which is one of the most common cause of obstructive uropathy leading to childhood renal failure. Most of the congenital uropathies are diagnosed antenatally, but in a developing country like India, late presentation is common due to ignorance and illiteracy. Hence these patients require corrective surgeries for which giving anaesthesia is a challenge. Present paper is an attempt to outline the anaesthetic management in these patients.

CASE REPORT

A 2 years old male child, weighing 10 kilogram(kg)was posted for bilateral ureteric implantation. He was a known case of craniosynostosis. Magnetic resonance imaging (MRI) of the brain at birth revealed premature fusion of sagittal suture, metopic suture predominantly on the right side with crowding of posterior fossa. Partial corpus callosal agenesis predominantly of tail and body was seen. There was no area of altered signal intensity or restricted diffusion. He had delayed milestones, could recognize parents, spoke monosyllables, had control over bladder and bowel but was unable to sit or stand on his own. Few days after birth, he underwent cystoscopy with posterior urethral valve fulguration. During first year of his life, he was advised exogenous steroids and stopped later on medical advice. He also received thyroid hormone supplements for first six months of life.

Repeat Magnetic Resonance Imaging (MRI) in March 2016 revealed a normal pituitary gland but thinning of posterior body of corpus callosum. Cerebral parenchyma, brainstem and cerebellum revealed normal signal intensity.

Dimercapto-succinic acid (DMSA) scan, few days after birth revealed normal cortical function of both kidneys with no evidence of scars or active infection.

Recent DMSA scan revealed scarred left kidney with mild to moderate impaired cortical function and scarred right kidney with preserved cortical function. Patient was now posted for bilateral ureteric implantation.

Micturating cystourethrogram revealed reflux on both sides in kidney and ureter. There was narrowing of vesico urethral junction. His thyroid hormones & cortisol levels were within normal range.

Airway examination revealed bad dentition and oral hygiene and a small mandibular length. Preoperative anesthetic plan was general anesthesia with endotracheal intubation with epidural analgesia. Hence spine imaging was done preoperatively to rule out spina bifida which is one of the associated abnormalities in these patients. Mother of the child was advised to do chlorhexidine mouth wash of the child for oral hygiene. In view of difficult airway video laryngoscope was made available.

On the day of surgery, written informed consent was taken from parents. 22 gauge intravenous (IV) cannula was in situ as patient had been on IV antibiotics. Electrocardiogram leads (ECG), pulse oximetry, non invasive blood pressure (NIBP) monitors were attached. Video laryngoscope with pediatric blade was kept ready. General anesthesia was induced with sevoflurane (4%), oxygen and air, IV propofol 15 mg, atracurium 5mg and fentanyl 10 microgram(mcg). Trachea was intubated with 4.5 uncuffed endotracheal tube and ventilation maintained with Jackson Rees modification of Ayre's T piece. Under aseptic precautions epidural catheter was placed through the caudal space and 5 ml of 0.125% Bupivacaine was injected. Surgery lasted for two hours. Intraoperative vital parameters were stable, blood loss was minimal. Trachea was extubated after neostigmine reversal. Postoperative Epidural Ropivacaine 0.1% infusion was started at the rate of 3ml per hour. It was given for 48 hours and patient was comfortable.

Post-operative course was uneventful, he was discharged on day 5 post surgery.

DISCUSSION

Agenesis of the corpus callosum (ACC) has an incidence of 0.5-70 in 10,000. There appears to be a male predilection (M: F ~2:1). Maternal alcohol consumption during pregnancy has been recognised as another risk factor.[2]

Partial absence is called dysgenesis. The corpus callosum is the major interhemispheric fiber bundle in the brain, consists of about 200 million axons making it the largest fiber tract in the central nervous system. ACC is a heterogenous condition which occurs as an isolated condition or as manifestation of a congenital syndrome. Rubinstein et al found that partial appearance of the corpus callosum may be due to a process to overcome initial abnormalities of midline structures resulting in a variety of shape, size and location of an observed callosal structure not necessarily corresponding to a normal corpus callosum.[3]

Agenesis of corpus callosum produces symptoms during the first two years of life in almost 90 % of patients. It was thought to be a rare condition but increased use of neuroimaging has increased rate of diagnosis. Currently incidence is 7 in 1000. Antenatal ultrasound can reveal this abnormality in utero. The Exogenous factors responsible for this condition are maternal alcohol use during pregnancy, maternal phenylketonuria or genetic factors.[4] Maternal history ruled out above causes in present case.

Perhaps, the best known metabolic disorder associated with this malformation is nonketotic hyperglycinemia, a condition typically associated with a neonatal encephalopathy. As many as 40% of patients with this disorder may have callosal agenesis, reflecting the fact that metabolic derangements may begin in early prenatal life, affecting fetal development. Another significant group of metabolic disorders associated with agenesis of the corpus callosum is the group of defects in pyruvate metabolism including pyruvate dehydrogenase and pyruvate carboxylase deficiencies. These disorders are typically associated with lactic acidosis and it is of interest that, in some cases, they are also associated with dysmorphic facial features. Present case had dysmorphic features but without any of above metabolic problems.

Clinical findings include mental retardation (60%), visual problems (33%), speech delay (29%), seizures (25%) and feeding problems (20%). It is known that patients with normal intelligence, mild behavioral or social problems and attention deficit hyperactivity disorder (ADHD) may have ACC.

Present patient had above problems, except seizures, though he was a case of partial absence involving tail and body. His karyotype was normal. He had undergone posterior urethral valve fulguration and now a bilateral ureteric implantation. He had been on intermittent urinary antibiotics since birth. His arterial blood gases were normal. A regular follow up was advised for his renal problems. He had deep set eyes, prominent forehead with progeric features and wrinkled skin. He was underweight and had hypotonia. He had a small mandibular length, hence video laryngoscope was kept ready. However, he did not pose any difficulty in intubation.

Spina bifida can occur with this condition, hence it was ruled out on imaging preoperatively. Hence, epidural anesthesia could be given in present case. His recovery was uneventful.

Special education, supportive care and physiotherapy are the mainstay treatment for this condition. Specific treatments like antiseizure medications, shunt surgery for hydrocephalus are done specific to the case. Genetic counselling may be of benefit to the families.

There is ongoing research being done on cognitive and psychosocial aspect and genetic studies of corpus callosal agenesis.

CONCLUSION

Present case shows that patients with Corpus callosal agenesis with craniosynostosis with pagiocephaly may have varied presentations & anesthetic implications like small mandible, spina bifida, etc. Preoperative imaging and planning is essential. Each case requires customization of anaesthetic techniques depending on the presentation.

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