Case Report

Anaesthetic management in a child with an atypical triad for reconstructive scoliosis surgery

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ABSTRACT

Scoliosis may be of varied aetiology and may be associated with severe congenital anomalies. It often poses a challenge in its anaesthetic management. We present anaesthetic management of a child who underwent scoliosis reconstruction with a rare triad of cerebral palsy, glucose-6-phosphate dehydrogenase deficiency and severe mitral regurgitation. Anaesthetic management in these patients should focus primarily on associated co-morbidities and congenital anomalies affecting the course of the perioperative management and thereafter comprehensive pre-operative strategies must be executed to enhance the safety profile during the surgery.

Key words: Cerebral palsy, glucose-6-phosphate dehydrogenase deficiency, haemolysis, mitral regurgitation, scoliosis

INTRODUCTION

Scoliosis is a complex deformity of spine characterised by lateralisation of spinal curvature with malrotation of vertebrae and mainly affects cardiovascular, respiratory and nervous systems.[1] Anaesthetic management of these patients have special concerns due to associated co-morbidities. We present the anaesthetic management of a child who underwent scoliosis reconstruction with a rare triad of cerebral palsy (CP), glucose-6-phosphate dehydrogenase (G6PD) deficiency and severe mitral regurgitation.

CASE REPORT

An 11-year-old, 26-kg female patient was admitted in orthopaedic unit with complains of backache, difficulty in walking and deformity of spine and was diagnosed case of G6PD deficiency and CP. She had severe thoracolumbar scoliosis (Cobb's angle - 55°) and was planned for surgical correction of the deformity. She had a history of premature delivery with low birth weight (1500 g) and epilepsy on oral carbamazepine for the last 5 years. On chest auscultation, bilateral coarse crepitations in lower zones and a pansystolic murmur in precordial area were heard.

Pulmonary function tests (PFT) and echocardiography with colour Doppler were advised, but the patient was not able to perform PFT. Echocardiography showed moderate pulmonary artery hypertension (pulmonary artery pressure - 32 mmHg) with left ventricular ejection fraction of 70%and grade-3 mitral regurgitation. Patient was scheduled for surgery after optimizing chest condition with full course of antibiotics and bronchodilators. On the day of surgery, infective endocarditis prophylaxis was instituted (amoxicillin 1 g PO and Inj.Gentamicin 40 mg 1 h prior to the surgery) in view of severe regurgitant lesion.

After obtaining written informed parental consent the child was transferred to the operating room and all baseline monitors (non-invasive blood pressure, continuous electrocardiogram, oxygen saturation-pulse oximeter and bispectral index) were attached. Child was pre-medicated with glycopyrrolate (125 μg IV) and fentanyl (60 μg IV). Anaesthesia was induced with propofol (60 mg IV) and muscle relaxation

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was achieved by administration of atracurium besylate (15 mg IV). Trachea was intubated with cuffed endotracheal tube and anaesthesia was maintained with rate controlled infusion of propofol (6 mg/kg/h, following the 10-8-6 regimen to achieve effective plasma concentration).[2] Atracurium (0.5 mg/kg/h) and fentanyl (1 μg/kg/h) with 50% O_2 in air. Left radial artery and ultrasound guided right internal jugular vein cannulation were carried out for continuous haemodynamic and central venous pressure monitoring respectively. Peripheral nerve stimulator was applied and urinary catheter and nasopharyngeal temperature probe were inserted. Appropriate measures to maintain normothermia were employed including warming blankets and warm fluids infusion. Intraoperatively, wake-up test was performed successfully. Tranaexemic acid (loading dose of 10 mg/kg just before surgical incision followed by continuous infusion of 1 mg/kg/h until closure of skin incision) was also administered to minimise intra-operative blood loss. Surgery lasted for nearly 7 h with approximate blood losses of 1200 ml, which was adequately replaced.

Post-operatively, patient was transferred to intensive care unit (ICU) for elective ventilation in view of hypothermia (32°C) following prolonged surgery, which was not responding to aforementioned warming measures. Patient was electively ventilated for next 24 h on continuous infusion of fentanyl (2 μg/kg/h IV) and dexmedetomidine (0.2 μg/kg/h IV). Trachea was extubated next day uneventfully. On day 3, patient was shifted to high dependency unit and subsequently discharged on the 7th day without any complication.

DISCUSSION

Anaesthetic considerations in scoliosis surgery need special attention due to the concerns pertaining to nature of surgical intervention and coexisting multiple diseases.[1] Severe scoliosis may itself affect nearly all physiological systems and its association with severe congenital anomalies may further increase the risk.[1]

Patient was a diagnosed case of spastic CP. The perioperative management strategies in these patients mainly focus on cardiopulmonary optimisation, post-operative pain management and active measures to prevent hypothermia and seizures.[3] Most of these patients are on anticonvulsants and thus epileptogenic agents such as ketamine, pethidine, tramadol, etomidate and enflurane should be avoided. Minimum alveolar concentration (MAC) values are altered in these patients particularly those receiving anticonvulsants, hence volatile agents should be used with caution.[3] We although opted for total intravenous anaesthesia (TIVA) in this patient and avoided volatile agents because of the possible association of malignant hyperthermia (MH) in these patients with scoliosis. Central vascular access may be difficult in these patients due to contractures and improper postures.[3] Ultrasound guided cannulations may be helpful and possibly avert the complications associated with conventional techniques. Post-operative pain management needs multimodal regimen and active counselling of patients because of depressed intellectual disability or poor verbal communication skills. Active warming measures should be initiated before induction of anaesthesia as these patients are particularly prone to hypothermia owing to extensive and prolonged surgery and their inability to regulate temperature.[3] Besides warming measures they may require post-operative care as observed in this child who was electively ventilated for 24 h. We prescribed a combination of dexmedetomidine and fentanyl infusion post-operatively in ICU. Dexmedetomidine (α2-adrenoceptor agonists) is a sedative, anxiolytic, sympatholytic with opioid sparing action and hence may be beneficial in these patients.[4]

G6PD deficiency was an incidental finding seen in this patient. On search of the medical literature we could not find any association of this disease with scoliosis, CP or valvular heart disease. Specific tests such as quantitative spectrophotometric analysis and rapid fluorescent spot test were not available in our institution for further classification. We presumed that the patient belongs to Mediterranean variant (World Health Organization Class 2), a common variant prevalent in Indian subcontinent.[5] Although most affected individuals are asymptomatic, but can develop severe haemolytic anaemia triggered by certain exogenous agents and drugs such as thiobarbiturates, sodium nitroprusside, diuretics, phenytoin and sulphonamides.[5] The pre-anaesthetic record should clearly mention all these drugs to avoid any catastrophes during intraoperative period. Both propofol and fentanyl are safer options. Acute haemolytic crisis in the perioperative period is difficult to diagnose as there are no immediate signs during anaesthesia. Hypotension is non-specific indicator and haematuria is a delayed sign. Maintenance of urine output is necessary and haemogram should be done daily in post-operative period.

Literature shows association of MH with myopathies in patients having scoliosis.[6,7] MH is one of the clinical entities triggered by depolarising muscle relaxant suxamethonium and nearly all volatile anaesthetic agents. We therefore adopted TIVA technique with atracurium infusion for the aforementioned reasons. There is a high incidence of post-operative neurologic injury after major reconstructive spine surgery.[8] Intraoperative neurological monitoring can improve patient outcome by allowing early detection of ischaemia before irreversible damage. Neuro-monitoring techniques to test spinal cord integrity during intraoperative period includes somatosensory evoked potentials, transcranial motor evoked potentials, continuous and triggered electromyography activity and Stagnara wake-up test.[8] Wake-up test has definite limitation as it is dependent on patient compliance and carries the risk of self-extubation, intraoperative awareness, loss of safe surgical positioning and providing only global assessment of spinal cord function.[8] Since other modes for neurological assessment were unavailable in our institution, we employed this technique. Blood loss is another concern in scoliosis surgery. Common strategies to minimise blood loss in perioperative period include cell saver, recombinant factor VIIa and anti-fibrinolytic agents (aprotinin, tranexamic acid, epsilon-aminocaproic acid).[9] We administered continuous infusion of tranaexamic acid during the surgical procedure with favourable results.

This child also had severe (grade 3) mitral regurgitation. Anaesthetic goals should be to keep heart rate high with low afterload to promote forward flow. Volatile agents cause vasodilatation and the resulting increase in heart rate is beneficial to regurgitant lesion but they should be used cautiously in these patients having altered MAC as it may depress contractility and may further worsen the regurgitant flow.

CONCLUSION

Anaesthetic management in these patients with this rare triad should focus primarily on optimisation of cardio-respiratory functions, prevention of perioperative hypothermia and management of post-operative pain and extensive blood loss. Anaesthetic drugs should be judiciously administered keeping in mind of the implications of G6PD deficiency and MH. Comprehensive pre-operative strategy and perioperative management of all coexisting diseases will enable a successful outcome during the surgery.

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