Anesthetic management in a case of Fraser syndrome

Sir,

Fraser syndrome is a rare syndrome with an autosomal recessive mode of inheritance. The main feature is cryptophthalmos (hidden eye). Other features associated are mental retardation, craniofacial dysmorphism, ear malformations, orofacial clefting, and malformation of the larynx, syndactyly, ambiguous genitalia renal malformation, and musculoskeletal anomalies.[1] We report a case of Fraser Syndrome and its anesthetic implications.

A six-year-old female child weighing 11 kg presented to the Ophthalmology Department for cryptophthalmos. Her language and fine motor developmental milestones were delayed. Her facial features were right eye cryptophthalmos and corneal opacity, left eye ptosis, ala nasi absent on the right side, triangular face [Figure 1],

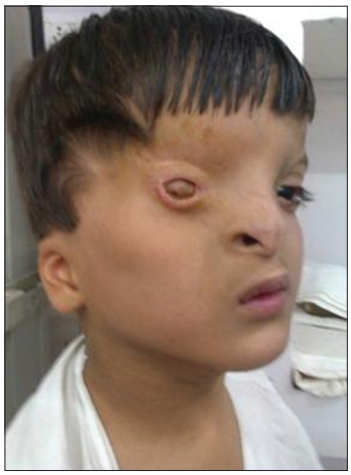

Figure 1: Facial appearance showing right eye cryptophthalmos and corneal opacity, left eye ptosis, absent ala nasi on the right side, triangular face, and low-set ears

high arched palate, tongue-tie, and low set ears. External genitalia were hypoplastic with no vaginal opening. On clinical examination of the chest and heart, no abnormality was found. Investigations were haemoglobin of 9 gm% and normal liver function test. Her two-dimensional echocardiogram was normal. The child was scheduled for symblepharon release surgery. On arrival to the operating room, standard monitoring, i.e., pulse-oximetry and electrocardiography were started. Anaesthesia was induced with tidal breathing of sevoflurane 7% with 100% oxygen. As anticipated, mask ventilation was difficult due to facial dysmorphism. An appropriate sized oropharyngeal airway was inserted to facilitate induction and ventilation of lung. After adequate depth of anesthesia was achieved, intravenous access was secured with a 22 G intravenous cannula. After administration of fentanyl 20 μg, IV and atracurium 5 mg IV, the trachea was intubated with a 5 mm uncuffed endotracheal tube. Anesthesia was maintained with isoflurane with O_2 and N_2O. The patient was mechanically ventilated to maintain adequate oxygenation and normocapnia. Perioperative course was uneventful.

The abnormal facial features are present in patients with this syndrome. The most commonly reported ones are broad nose with depressed or flat nasal bridge, malformed or low set ears, microtia, atresia or stenosis of the external auditory meatus, and high arch palate. Clefting of lip/palate, laryngeal stenosis, and laryngeal atresia may also be present in these patients.[1,2] These facial anomalies can make mask ventilation, laryngoscopy, and laryngeal mask airway insertion difficult.[2] Awake intubation by fiberoptic bronchoscope or direct laryngoscopy along with the aid of a gum elastic bougie or an illuminating stylet is a possible option. Various supraglottic airways present a useful alternative to endotracheal intubation or can facilitate endotracheal intubation.

These patients may present with atrial and ventricular septal defect, dextrocardia, univentricular heart, a variant of Ebstein anomaly, coarctation of the aorta, and transposition of the great vessels.[3,4] Meningoencephalocele and spina bifida occulta are some of the common neurological anomalies associated with Fraser syndrome. Other less commonly reported abnormalities are hydrocephalous, encephaloceles, mild cerebellar hypoplasia, and periventricular leukomalacia. Renal agenesis with or without ureter agenesis or cystic dysplasia of the kidney and abnormalities of the urinary tract may present with cryptophthalmos.[3,5]

The patient with Fraser syndrome may be scheduled for ophthalmic, cosmetic, and genitourinary surgeries. Involvement of multiple systems can pose considerable challenges to the anesthetist. Careful planning and perioperative management are essential for successful anesthetic management of these patients.

Virender Kumar Mohan, Aanchal Sharma

Department of Anesthesiology, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence:
Dr. Virender Kumar Mohan,
Department of Anaesthesiology, All India Institute of Medical Sciences, New Delhi, India.
E-mail: dr_vkmohan@yahoo.com

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