Anesthetic Management of a Case of Robinow Syndrome

Sir,

Robinow syndrome consists of mesomelic brachymelia, short stature, genital hypoplasia, and a characteristic facies.[1] It is also known as fetal facies syndrome as the facial features of infants with this syndrome resemble those of an 8-week-old fetus.[2] Both autosomal dominant and recessive modes of inheritance are known for it with the recessive form having more severe symptomology.[2] It has an incidence of 1:500,000 and a 1:1 male:female ratio.[3] Cases have been reported from the Arab countries, Czechoslovakia, and the Indian subcontinent.[2] There are very few reports regarding the anesthetic management of Robinow syndrome.

A 6-year-old boy with Robinow syndrome having congenital hypospadias was posted for an elective urethroplasty. The child weighed 25 kg was alert, cooperative, of normal gait and had a normal height and limb length. He exhibited a characteristic facies with frontal bossing, ocular hypertelorism, wide palpebral fissures with an antimongoloid slant, and relatively large eyes. He had a depressed nasal bridge, an elongated philtrum, right syndactyly between the fourth and fifth fingers, left syndactyly between the third and fourth fingers, and micropenis [Figure 1].

His laboratory investigations were within normal limits. Two-dimensional echocardiography, abdominal ultrasonography, and spine and chest radiographs were normal. He was accepted under the American Society of Anaesthesiologists Grade II risk and general anesthesia was planned.

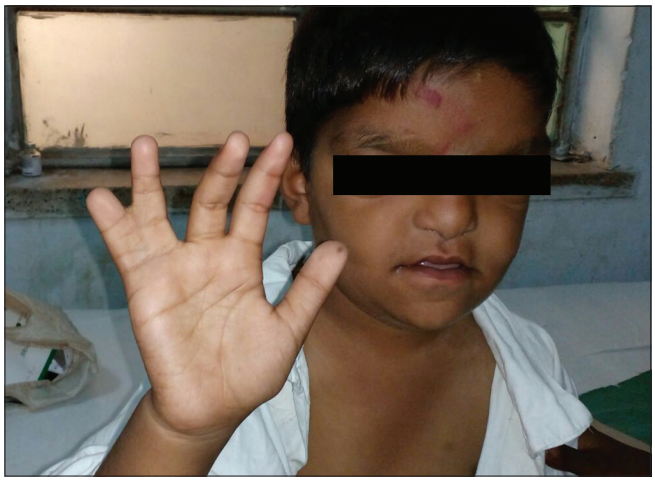

Figure 1: Depressed nasal bridge, an elongated philtrum, right syndactyly between the fourth and fifth fingers

The difficult airway cart was kept ready. Standard monitors were attached. He was premedicated with intravenous (IV) glycopyrrolate 0.2 mg, midazolam 1 mg, and fentanyl 50 mg on the operation table. After preoxygenation, he was induced with injection propofol 50 mg. Mask ventilation was possible. IV succinylcholine 50 mg was administered. Laryngoscopy revealed a grade one Cormack–Lehane view of the glottis. He was intubated with 5.5 size cuffed endotracheal tube and maintained with nitrous oxide, oxygen, sevoflurane, and atracurium. At the end of the procedure, he was reversed with neostigmine 1 mg and extubated. The postoperative course was uneventful.

Autosomal dominant Robinow syndrome is characterized by skeletal findings such as short stature, mesomelic limb shortening predominantly of the upper limbs, and brachydactyly; genital abnormalities such as micropenis, webbed penis, cryptorchidism, hypoplastic clitoris, hypoplastic labia majora, and dysmorphic facial features, especially in early childhood; dental abnormalities such as malocclusion, crowding, and hypodontia; vertebral abnormalities such as hemivertebrae and scoliosis; cardiac defects, more commonly pulmonary stenosis and pulmonary atresia, cleft lip/palate.[4]

Autosomal recessive Robinow syndrome is characterized by distinctive craniofacial features, skeletal abnormalities, and other anomalies.[5]

The diagnosis mainly depends on the clinical features.[2] Our patient had most of the clinical features of the autosomal dominant variety.

They usually need corrective surgery for limb and spine defects, facial abnormalities, orthodontic problems, vaginal atresia, hematocolpos, scrotal transposition, and umbilical hernias.[4,5] We chose general anesthesia technique as the child was uncooperative, the parents were sceptical of regional blockade, and the procedure was of long duration.

Facial dysmorphic features such as midface hypoplasia, gum hypertrophy, cleft palate, and crowded teeth can make the airway difficult in these patients.[1,2,4,5] Some authors have reported the occurrence of intrathoracic tracheomalacia during anesthesia in a case of Robinow syndrome.[1] In our case, although the face appeared abnormally shaped, the airway management was uneventful. Spinal or epidural technique can become difficult because of hemivertebrae. Associated cardiac defects can make the perioperative management very challenging. Preoperative radiological evaluation for vertebral and rib anomalies, upper airway obstruction or defects, and renal and cardiac function is recommended before sedation or anesthesia in patients with Robinow syndrome.[6]

Our case depicts that careful preoperative evaluation, planning, and appropriate technique with anticipation of a difficult airway and a difficult spine are must for the successful outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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