Anaesthetic challenges in a patient with acromesomelic dysplasia posted for vitreoretinal surgery - A case study

Dear Editor,

Acromesomelic dysplasia is a rare type of osteochondrodysplasia which is characterised by acromesomelia (unusual shortening of bones of arms, forearms, legs, hand and feet), camptodactyly (bent fingers), bilateral clubbed feet, short stature, scoliosis and flattened midface.[1] These cases pose a challenge to anaesthesiologists due to the anticipated difficult airway, obstructive sleep apnoea, pulmonary arterial hypertension, scoliosis and restrictive lung disease, thereby predisposing to higher postoperative pulmonary complications and joint dislocations while positioning etc.[2]

A 34-year-old, 20-kg male with acromesomelic dysplasia was scheduled for right eye vitreoretinal

surgery for retinal detachment. On pre-anaesthetic examination, he had the characteristic phenotype of the syndrome with upper and lower limb deformities [Figure 1]. He was bedridden and had normal intelligence. Airway examination in the supine position revealed a 5 cm mouth opening with Modified Mallampati Class II, restricted neck movements and abnormal dentition (spaced teeth) whereas blood investigations, electrocardiogram and echocardiography were normal, but chest radiograph revealed scoliosis from D-2 to L-2 vertebrae (Cobb's angle of 60°) with convexity to the right side [Figure 2]. A pulmonary function test could not be done as the patient could not perform spirometry due to his inability to sit. His heart rate was 80/min, and blood pressure was 138/88 mmHg. The patient was informed about the difficult airway, high risk of postoperative pulmonary complications and mechanical ventilation due to the severity of scoliosis with general anaesthesia. The patient was also explained about the option of surgery under regional anaesthesia. The patient declined consent for general anaesthesia and gave consent for regional anaesthesia. The patient was shifted to the operation theatre on the day of surgery. Bony prominences and pressure points were padded while positioning the patient to avoid any neurological, skeletal injuries or joint dislocations. Monitors (electrocardiogram, blood pressure and pulse oximetry) were attached, and a 20-gauge intravenous cannula was secured in the left hand. Oxygen was supplemented using nasal prongs with 2 L/min of flow with continuous end-tidal carbon dioxide ($EtCO_2$) monitoring. A bolus of intravenous dexmedetomidine 1 μg/kg was administered over 10 min, followed by an infusion of 0.5 μg/kg/h. Topical proparacaine 0.5% was instilled in the operative eye, followed by a sub-tenon block with a 5 mL mixture of 2% lignocaine and 0.5% ropivacaine administered by the ophthalmologist. The patient did not report pain while administering the sub-tenon's block and surgery. The duration of surgery was 90 min, and vitals remained stable. Paracetamol 15 mg/kg was administered intravenously 10 min before and dexmedetomidine infusion was stopped at the end of the surgery. The patient was transferred to the post-anaesthesia care unit uneventfully.

Acromesomelic dysplasia is a rare form of dwarfism. Our patient was non-ambulatory due to his phenotype. We took care during positioning of the patient on the operating table and during shifting due to the propensity of joint dislocation. Regional anaesthesia should be preferred to avoid the risks associated with general anaesthesia. However, there may be technical difficulties due to possible anatomic variations. Local anaesthetic (LA) agent dose should be carefully calculated to prevent overdosing and toxicity, as these patients weigh less. Our patient's weight was only 20 kg, so we planned to administer a sub-tenon block over the peribulbar block as it requires only 5–7 mL of LA and is less painful with fewer complications. A difficult airway cart was also kept due to an anticipated difficult airway along with an intensive care unit arrangement, given the high risk for postoperative pulmonary complications due to scoliosis, restrictive lung disease and restricted neck movements. Small-sized endotracheal tubes were kept as age-related endotracheal tube size prediction is reported to be difficult.[3] In the present case, dexmedetomidine provided conscious sedation and analgesia during the subtenon's block and surgery.[4] Dexmedetomidine has been used in syndromic patients.[5] Recently, dexmedetomidine has

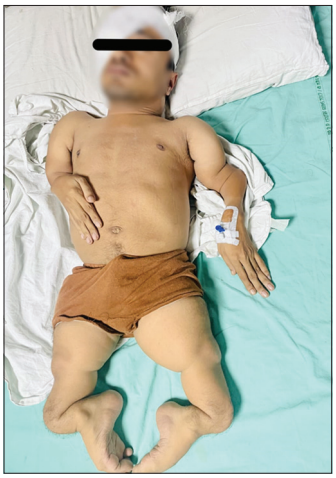

Figure 1: Patient with acromesomelic dysplasia phenotype

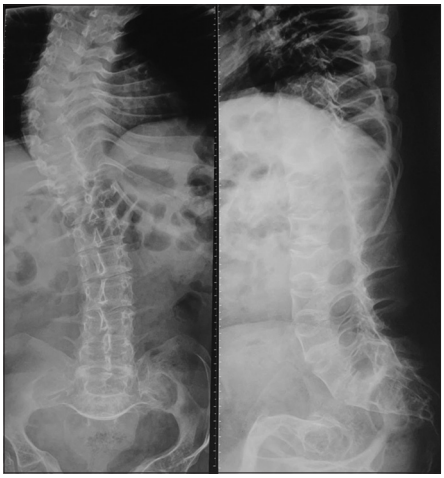

Figure 2: Spine X-ray showing dorsolumbar scoliosis

been used in patients with mitochondrial disorders as a safer alternative to other anaesthetic agents (propofol/inhalational agents).[6]

To conclude, patients with acromesomelic dysplasia and other syndromic short stature require particular caution due to anticipated difficult airways. Regional anaesthesia can be administered with careful assessment of the dose of LA to avoid toxicity. Adjuvants such as dexmedetomidine can be administered for anxiolysis and sedation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient consented to his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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