Perioperative management of adult patient with congenital varicella syndrome for oncologic surgery

Sir,

Varicella-Zoster virus can cause severe complications in immunocompromised individuals. We encountered a 28-year-old, unmarried female, posted for a Right Modified Radical Mastectomy with Axillary clearance.

She was 136 cm tall, weighed 25 kg. She had cicatricial lesions on the right side of her face. Her right eye was affected. It was smaller, with a cloudy cornea, only perception of light and inability to close that eye completely. Her left hand showed polydactyly and both lower limbs had syndactyly [Figure 1]. 2D Echo which was done, since the patient had completed 12 cycles of Paclitaxel and 4 cycles of Cyclophosphamide and Epirubicin, showed ejection fraction of 55% with a jerky interventricular septum. She understood simple commands. Airway examination revealed deformed nostrils; multiple loose and missing teeth, in upper and lower jaw, Mallampati grade I, 4 cm mouth opening, restricted neck extension, sternomental distance of 8 cm and thyromental distance of 4 cm. 22-gauge intravenous cannula was secured in the left arm after three attempts. Anticipating a difficult airway, after pre-oxygenation, patient was induced with 50 µg of Fentanyl, 50 mg of Propofol and 50 mg of Succinylcholine. The patient was intubated in the second attempt, after changing from Macintosh 2 to Macgrath Mac 3. Cormack-Lehane improved from 3b to 2b following which, a number 6.5 cuffed portex tube was inserted using a stylet. Standard ASA monitoring was employed. The patient had an uneventful intra-operative course and was successfully extubated at the end of surgery.

Pregnant women contracting Varicella between 7 and 28 weeks of pregnancy may infect the foetus and lead to Congenital Varicella Syndrome (CVS)(0.4%–2.2%).[1] CVS is usually characterised by low birth weight, cutaneous scars in dermatomal outline (70%), papular lesions, ocular abnormalities (66%), CNS abnormalities (46%) and poor sphincter control (32%).[2] The characteristic scarring that is seen represents cutaneous residua of the VZV infection of the sensory nerves. Tissues in a rapid developmental stage, e.g., limb-buds, are affected, resulting in shortened and/or malformed extremities. CVS has 30% mortality in infancy.[3] Our patient had cutaneous scars restricted to the right side of the body, her right eye was affected, showed delayed milestones and had malformed extremities.

There is a paucity of CVS literature, with only about 130 cases reported, most from paediatric population.[2] Hence, the challenges faced, either in administering anaesthetics or peri-operative behaviour of adult patients undergoing surgery remain undocumented. We faced multiple challenges which included a difficult venous access, a difficult airway, difficulty in obtaining consent and ocular deformity leading to the possibility of corneal injury.

Securing a venous access was difficult, which was concurrent with the findings of Bensghir et al. that identified multiple cycles of chemotherapy as an independent risk factor for difficult venous cannulation.[4]

Mental retardation is a known feature of CVS,[2] which led to difficulty in explaining the procedure to the patient and allaying her anxiety. The consent was taken from her brother.

The small stature and facies, multiple missing teeth and restricted neck movements prognosticated a difficult airway. She needed a smaller mask and blade size, and when conventional laryngoscopy failed to

Figure 1: (a) Multiple loose and missing teeth, cicatricial scarring. (b) Inability to close the right eye completely. (c) Syndactyly in lower limbs. (d) Polydactyly in upper limbs
give us a good view, we used videolaryngoscopy as per the ASA difficult airway guidelines.\[6\]

Since the patient was unable to close her damaged eye, we used methylcellulose eye drops and eye padding to prevent further damage.

Considering the high morbidity and mortality, in paediatric population and afflicted foetus, immunising children and women of childbearing age against Varicella is a good strategy. Vaccination is not recommended during pregnancy. Prophylactic administration of zoster immunoglobulin (ZIG), within 96 hours of exposure can prevent or reduce the severity of varicella.\[2\]

The management of our case was unique because we had no available literature to anticipate perioperative outcomes. Our case adds to the limited literature on CVS and its anaesthetic management.

**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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