Anesthetic management of a patient with Rosai-Dorfman disease and atrial septal defect for orbital surgery

Sir,

Rosai-Dorfman disease (RDD) is a rare histiocytic disorder characterized by the presence of sinus histiocytosis with massive lymphadenopathy involving mainly cervical and at times mediastinal, inguinal and retroperitoneal nodes.[1] Extranodal involvement is rare but may affect the skin, soft tissue, upper respiratory tract, bone, eye, urogenital tract, breast, gastrointestinal tract, liver, pancreas, thyroid and lungs.[2-5] Systemic steroids, alkylating agents and radiotherapy are used to control the disease. Surgery is generally limited to biopsy, but debulking may be required in patients with vital organ compromise. Multisystem involvement and the airway in particular are the usual anesthetic concerns. Serious problems in form of airway obstruction, superior vena cava syndrome or cardiopulmonary collapse may occur if the trachea or vital structures in the mediastinum are compressed. We report the anesthetic management in a patient with RDD who underwent debulking surgery of the orbital tissue.

An 18-year-old, 48 kg, woman presented for debulking of a right-sided periorbital swelling. The diagnosis of RDD was made on lymph node biopsy and patient was not on any medication. Patient had a history of snoring though there was no history suggestive of obstructive sleep apnea. Apart from a soft, non tender periorbital swelling, she had preauricular nodes on the right side and bilateral cervical and submandibular lymphadenopathy [Figure 1]. Her airway was adequate with no signs of compression. Contrast enhanced computed tomography (CECT) of the neck, chest and abdomen revealed cervical lymphadenopathy, but no mediastinal or abdominal lymphadenopathy. The pulmonary artery was prominent in the CECT and echocardiography showed an atrial septal defect (ASD) 1.5 cm with left to right shunt. There was no pulmonary hypertension.

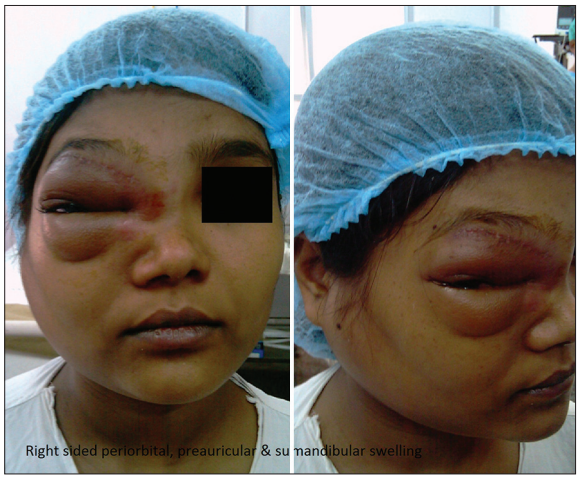

Figure 1: Front and side view of patient's face showing facial asymmetry

A difficult airway cart was kept standby in the operating room. After applying standard monitors and securing an intravenous (IV) access, 1 g of paracetamol was infused over 20 min. The right eye was covered with a sterile eye pad to prevent any injury. The patient was pre-oxygenated with 100% oxygen. As anticipated, mask holding was difficult because of the orbital and pre auricular swelling. Adequate mask ventilation was achieved by using an oral airway and using both hands to get the mask seal. Anesthesia was induced by tidal breathing of sevoflurane 7% in a mixture of oxygen and air. Once adequate depth was achieved, a Laryngeal Mask Airway (LMA) Proseal (Size 3) was inserted to secure the airway. A 12 Fr gastric catheter was placed in the drain tube of the LMA to evacuate air from the stomach, which had entered due to difficult mask ventilation. Fentanyl 100 mcg was administered IV after securing the airway. The lungs were mechanically ventilated using pressure support mode (PS: 18 hectapascal, Flow 1 l/min, Trigger 0.3 L/min). The LMA had to be manipulated to achieve adequate ventilation (delta Vt: 34 ml, $EtCO_2$ 34-36 mmHg). Anesthesia was maintained with oxygen, air and sevoflurane. Heart rate and mean blood pressure were within 20% from their baseline values. There was no hypoxia, hypothermia or hypercarbia. Surgery proceeded uneventfully and the LMA was removed once the patient was fully awake. The patient was transferred to the post-anesthesia care unit for observation and monitoring of vital parameters and transferred to the ward after 6 h.

Multisystem involvement requires a thorough evaluation in these patients with particular emphasis on the airway. This may cause anxiety to these patients as frequent visits to various departments may be required. Communication and reassurance is therefore very essential. A history of snoring, sleep-apnea, seizures should be taken. The side effects

pertaining to long-term steroids and alkylating agents should be kept in mind during the perioperative period. Apart from X-ray of the neck, radiology workup should include a computed tomography (CT) of the head, neck, chest and abdomen to ascertain/exclude nodal and extranodal manifestations and possible pressure effect on airways. Our patient had a history of snoring along with bilateral cervical lymphadenopathy. However, there was no airway compression. Difficulties with ventilation and intubation should be anticipated. Mask ventilation may be difficult as was in our case. A difficult airway cart should be kept in the operating room. In case of suspected airway compromise, awake fiberoptic guided endotracheal intubation in a spontaneously breathing patient is the best possible option in these patients. Either a supraglottic airway or an endotracheal tube can be used for securing the airway, though the former is better tolerated by the patient when using an inhalational agent. We used LMA Proseal since it is a preferred supraglottic device in case of controlled ventilation.

We ensured hemodynamic stability by careful monitoring of the concentration of inhaled sevoflurane and fluid replacement as the patient had an ASD. Care of the eyes should be taken during mask ventilation. For monitoring of temperature, nasopharyngeal probes should be avoided, since the nose is a common site of extranodal involvement in RDD.[6] Care should be taken during positioning as RDD may be associated with the osteolytic skeletal lesions.

To conclude, it is essential for anesthesiologists to be aware of RDD and to carefully evaluate patients with this rare disease and assess the degree of organ involvement with special emphasis on the airway. Neuromuscular blocking agents may produce airway collapse and thus should be avoided.[7] Inhalation anesthesia with spontaneous ventilation is strongly recommended. A supraglottic airway like LMA is a safe option in these patients.

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