Airway management and anesthesia in posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects and eye abnormalities syndrome: A case with laryngotracheal hemangiomas

Sir,

Posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects and eye abnormalities (PHACE) is a rare neurocutaneous syndrome that was described by Frieden *et al*. in 1996.[1] Hemangioma

was reported in approximately 50% of the patients and it was located in the subglottic region in the majority (83%) of the patients.[2] The majority of the patients with PHACE syndrome need vascular, neurological, ophthalmologic or otolaryngologic surgery in their early ages.[3] According to the best of our knowledge, the anesthetic management of a patient who had airway hemangioma and PHACE syndrome has not been reported previously.

A 42-month- old (weighing 15 kg and height 101 cm) female with previously diagnosed PHACE syndrome was referred to the plastic and reconstructive surgery service for congenital blepharoptosis correction. A detailed medical history was recorded from patient's parents and available medical records. She was evaluated by the pediatric chest clinic after a stridor complaint when the patient was 12 months and a magnetic resonance imaging (MRI) angiography revealed agenesis of the right internal carotid artery and hemangiomas involving the left parotid gland and laryngotracheal space. Echocardiography showed that the intracardiac and aortic arch anatomy was normal and assessment with MRI did not indicate any posterior fossa abnormalities. The patient received propanolol and intermittent systemic steroid treatment for 24 months, there was a reduction in the size of the hemangioma and there was an improvement in the symptoms. Neuropediatric consultation revealed that she had normal motor and physical development and there were no cranial nerve anomalies. She had cutaneous hemongiomas along face and neck [Figure 1a]. The remainder of the physical examination and laboratory evaluation was normal.

After the application of routine monitors with BIS monitoring (Covidien), anesthesia was induced by the inhalation of sevoflurane and thiopental (4 mg/kg) with alfentanil (10 μg/kg) intravenously. Face mask ventilation was easy. Anesthesia was maintained with sevoflurane (1-2%) and remifentanil (0.1 μg/kg/min). No muscle relaxant was administered. A detailed examination with a flexible fiberoptic (FO) scope (Karl Storz, external diameter 2.8 mm) ruled out any oropharengeal hemangiomas before insertion of laryngeal mask airway (LMA). A lubricated size 2 Supreme™ LMA (Maidenhead, UK) was smoothly inserted and the cuff was inflated with 10 ml air. The FO scope was passed through the LMA for the examination of lower airway. According to the FO evaluation, there were hypervascular lessions on the mucosa of the proximal to the fourth tracheal wall and larynx [Figure 1b]. If necessary, we had planned to place the endotracheal tube (ETT) cuff above the carina and under the 4 tracheal ring. We made necessary preparations, as tracheostomy was another option. The operation was completed in

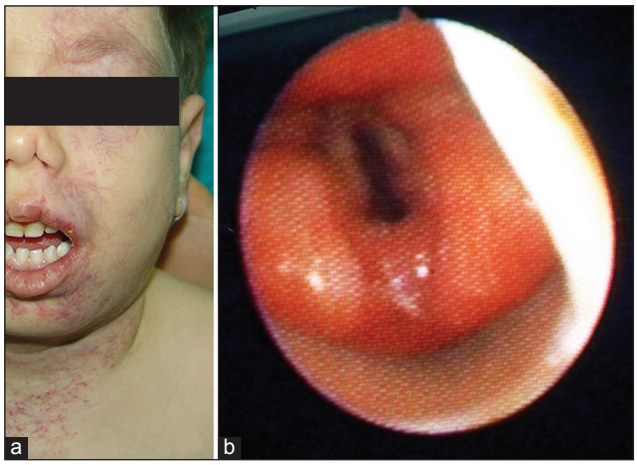

Figure 1: (a) Cutaneous hemongiomas along her face and neck. (b) Hypervascular lessions on the mucosa of the larynx

70 min without the need for ETT placement. There were no complications during the procedure. After withdrawal of LMA, we confirmed no bleeding inside the airway. Following operation, circulatory dynamics and respiratory status were stable and the postanaesthetic course was uneventful.

Airway hemangiomas can be a life-threatening aspect of PHACE syndrome. In the case series presented by Durr *et al*., 52% of the patients with PHACE syndrome had airway hemangioma and 25% of these patients required tracheostomy due to severe airway obstruction.[2] The symptoms associated with airway hemangioma were noisy breathing, difficulty breathing and stridor and approximately one-fifth of the patients were asymptomatic. The size of the hemangiomas, which are recovered after medical treatment, may increase later.[3,4] Therefore, we recommend performing airway evaluation in all symptomatic or asymptomatic PHACE patients before operation. Depending on the localization or narrowness, if present, ETT placement may be a difficult task. Moreover, ETT or cuff may exert direct damage to the hemangioma and cause edema, intratracheal bleeding and difficult ventilation. If ETT is absolutely needed, tubes with small number (without the use of stile, if possible) may be used after airway evaluation.[5] In this case, LMA did not exert any damage to the hemangioma, and allowed the observation of the laryngotracheal region using FOs.

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