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Anesthetic approach to a patient with Larsen syndrome

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Introduction: Larsen syndrome is a connective tissue disorder (LS), that may be both autosomal dominant and recessive and sporadic cases may also occur. This syndrome is characterized by large joint dislocations, spinal deformities, flattened nose bridge, hypertelorism, protruding forehead, and head and foot deformitiees and may be accompanied by pulmonary and cardiovascular anomalies. In this report, anesthetic apprach in a case of Larsen syndrome is presented.

Case: In a male patient at the age of 4 and weight of 14kg, orthopedics clinic planned to carry out operation for the relaxation of bilateral hamstring vs. gastrocnemius muscles. In physical examination, bilateral foot deformity, short neck, flattened nasal bridge, protruding forehead and hypertelorism were observed. No pathology was found in routine biochemical and hematological investigations. Vital findings were stable. Patients underwent routine monitorization with ECG, heart rate and pulse oximeter. Following approximately 5 minutes of premedication, anesthesia induction was carried out with 3mg/kg propofol and 2mcg/kg fentanyl. Considering that difficulty may be encountered in airway due to syndromic typical facial appearance, different sizes of face masks, airways and laryngeal masks were prepared for laryngoscopy, blades and endotracheal tubes at different sizes were kept ready and premedication was not performed. No problem was experienced in ventilation with face mask. In order to provide intraoperative-postoperative analgesia after induction, and to increase patient comfort, spinal block was made with 27G spinal needle. Mask ventilation was observed to be comfortable and 2,5 no I-Gel was placed without using neuromuscular blockers. Anesthesia maintenance was made with 50/50% azotprotokside/oxygen and 2%sevofluran mixture. Following the operation lasting for approximately 30 minutes, patients was extubated without any problems and patients was followed for 45 minutes in recovery room before being sent to clinic.

Discussion: Patients with Larsen syndrome may frequantly undergo operations for orthopedics and plastic surgery anomalies and due to these anomalies, their anesthesia has special features. Some difficulties are encountered in providing airway to patients with Larsen syndrome. In our case, neuromuscular blockers were avoided in consideration of probable difficulty of intubation and masks, tubes, blades at every size and LMA were prepared and a reliable and safe anesthesia was provided with spinal anesthesia and sevoflurane-N2O maintenance.

Conclusion: Patients with connective tissue disorders and syndromic ones are candiates for difficult intubation due to their primary diseases and restriction of movement. It is our suggestion that in Larsen syndrome, belonging to this group, complications may be decreased by providing optimal conditions in preoperative period, and in the induction and maintenance of anesthesia and by using spinal anesthesia along with general anesthesia.

Biography

Ozkan Onal has completed his medical education at Gazi University Medical Faculty and he was specialized in anesthesiology in Hacettepe University Medical Faculty. He has more than 15 publications in reputed journals in the field of anesthesia.

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