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Anestesia management in juvenile hyaline fibromatosis (JHF-Murray Puretic Drescher Syndrome)

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Introductıon: Juvenile Hyaline Fibromatosis (JHF-Murray Puretic Drescher Syndrome) JHF(juvenile hyaline fibromatosis, Murray Puretic Drescher Syndrome) is an autosomal recessive disease,characterized by growth retardation, gingiva hypertrophy, joint contractures and skin lesions. In its pathogenesis, collagen synthesis distrurbance plays role and it usually occurs in children. The most critical problem for anesthesia management is keeping the airways open in this report, airway management and use of frova intubation catheter as an option in a case of JHF is discussed and presented.

Case: A 13 year old male JHF case at the weight of 30 kgwas planned to undergo operation forth excision of soft tissue masses in nose tip, right-left ear posterior region and left temoral region on scalp. In physical examination, contracture was detected inneck, temporomandibular and extremity joints. All biochemical and hematological tests were normal. Routine monitorization, including ECG,non invasive blood pressure monitorization and pulse oximetry was carried out. Mallampati classification was 4 and preparations for difficult intubation were made. Following preoxygenization, induction was made with 20 mg lidocain,150 mg propofol and 100 micrograms fentanyl. Mask ventilation was comfortabale and rhocuronium 20mg was administered. Cormack-lehane classification was 4 and using frova intubation catheter, oral intubation was performed with no.6 endotracheal tube. In anesthesia maintenance, 250 mg/hour propofol and overall 3mg morphine was used. No problem was encounted during operation lasting 120 minutes. Heart rate and blood pressure were found to be stable between 10% lower and upper limits of preoperative values. Propofol infusion was withdrawn with the end of operation. Spontaneous respiration was adequate and patients was extubated without any problems.In follow up, no problem was encountered and the patient was discharged on 1st day postoperatively.

Discussion: JHF is arare autosomal recessive disease that arises usually at within the first few months of life. It is characterized by excessive accumulation for hyalin in many tissues such as skin, stomach-intestinal system, heart muscle, surrenals, skeletal muscles, spleen, lymph nodes thyroid tissue.Gingival hypertrophy is very widespread and characteristic of the disease. Tracheal intubation is difficult owing to gingival hypertrophy, and cervical spine and temporamandibular joint contractures.

Conclusion: It is our opinion that in JHF cases, preparations should be made for difficult airway in anesthesia management and that in such cases frova intubation catheter can be safely used.

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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