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Anesthetic approach to a case wıth familial Fahr syndrome

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Introduction and aim: Fahr syndrome (FS); is arare disease in which calcium and other minerals are deposited bilaterally and symetrically in basal ganglions, cerebellar dentate nucleus and subtantia alba. Its clinical findings are, parkinsonism,dystonia, tremor,chorea,ataxia, dementia, mood disorders, difficulty in swallowing, muscle cramps, seizure and speech disorders. In this report, our anesthesia experience in case of Fahr syndrome that was planned to undergo bilateral achiloplasty operation is presented.

Case: A 7 year old boy weighing 9 kg and was diagnosed with Fahr syndrome at the age of 1,5 was planned to undergo achiloplasty by orthopedics clinic due to contraction in bilateral extremities and unbalanced gait. In physical examination, speech, balance and gait disorders were present. Routine laboratory analysis results were normal.Two siblings of the patients were diagnosed also with Fahr syndrome and lost due to this disease. In operating room, blood presure was 110/65 mmHg, heart rate 104 beat/min, and SpO_2 was 98%,mallampati score was 2, and ASA score also 2. Preparation was made for difficult intubation with various size masks, tubes, blades and lma. In addition to standard monitorization, body temperature and BIS (bispektral index skala) monitorization was carried out. And dantrolen was prepared. 1 mg/kg lidocain HCI and 2 mg/kg propofol was administered. Patient was ventilated with 100% O2 and inserted 2,5 no i-gel. Anesthesia was maintained with 40% O2 and 60% N_2O mixture and 2mg/kg/h propofol infusion patient was hemodymanically stable and was extubated at the end of the operation. Then, he was transferred to orthopedics clinic with open consiciousness and well general condition.

Discussion: Metabolic disturbances, most of which are associated with calcium metabolism, occur in FS.Intracerebral calcifications usually accompany disturbances in serum calcium and phosphore metabolism. Due to these metabolic disturbances, anesthesia management should be planned, taking the risk of malignant hyperthermia into account. In addition, the probability of difficult intubation owing to calcium metabolism disturbance, should be borne in mind.

Conclusion: For early diagnosis of malignant hyperthermia due to volatile anesthetics and cardiac effect, careful monitorization, invasive blood pressure follow up and blood gas investigation should be carried out.As personality changes and impairment in mental functions is marked in Fahr syndrome and the etiology of intracerebral calcification can not be identified, we believe that general anesthesia is safer. In these patients, preparations should be madefor difficult intubation and care should be taken for the risk of the development of malignant hyperthermia in association with inhaler anethetics.

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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