Anesthesiological approach to a case with FAHR syndrome

Sir,

Fahr syndrome, defined by German neurologist Karl Theodor Fahr in 1930, includes neurologic, psychiatric, and cognitive disorders of unknown etiology, and is characterized by symmetric calcifications in the basal ganglia and cerebellar dentate nucleus, in individuals aged 5-65 years.[1-4]

A 58-year-old male patient, scheduled for inguinal hernia repair, had extrapyramidal movement disorders, speech-behavior disorder, urinary incontinence, a serum Ca level of 5.8 mg/dL, and diffuse calcifications in the cerebellum, basal ganglia, periventricular areas, and the occipital lobe gyri. Peripheral oxygen saturation, electrocardiography, and non-invasive blood pressure were monitored. Following anesthetization with 1 mg/kg^{-1} lidocaine, 50 μg fentanil, 2 mg/kg^{-1} propofol, and 0.6 mg/kg^{-1} rocuronium, the patient was ventilated with 100% O_2 and intubated orotracheally. Anesthesia continued with 50% O_2-50% N_2O in 2% sevoflurane. Arterial catheterization monitored ionized calcium levels and blood pressure invasively. During the operation, 10% calcium gluconate (20 mL) was administered by titration. Four minutes from the cessation of anesthetic inhalers, the patient opened his eyes. Muscle relaxant antagonization was not required and the patient was extubated.

Since, emergent interventions might be necessary in cases with a history of epileptic seizures,[5] general anesthesia was selected for this patient. Titrated calcium replacement therapy is recommended by monitoring ionized calcium levels, since calcium takes part in muscle contractions-relaxations.[6] Although, hypocalcemia affects neuromuscular agent metabolism, additional neuromuscular agents were unnecessary and no residual effects were observed. For Fahr syndrome patients, calcium levels should be closely monitored in cases with hypocalcemia, and general anesthesia should be used.

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