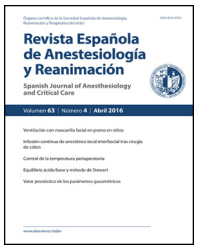
Rev Esp Anestesiol Reanim. 2016;xxx(xx):xxx–xxx

Revista Española de Anestesiología y Reanimación

www.elsevier.es/redar



CASE REPORT

Anesthetic management in a child with moya-moya disease and sickle cell anemia: Case report

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Received 1 December 2015; accepted 6 April 2016

KEYWORDS
Moya-moya disease;
Anemia;
Carotid stenosis

Abstract
Objective: The objective of this case report is to describe the anesthetic management in a child with moya-moya disease and sickle cell anemia provided in a tertiary hospital.
Case: A 6 year-old patient, diagnosed with moya-moya disease and sickle cell anemia, both conditions associated with a greater incidence of intracranial ischemic events, with a history of two strokes of the ischemic subtype, was submitted to general anesthesia for the execution of multiple cranial burr holes in order to produce the neovascularization in poorly perfused regions. There were no complications in the perioperative period and the child was discharged from the hospital on the second postoperative day.
Conclusion: Although scarcely described in the medical literature, the anesthetic management in a patient with moya-moya disease must ensure the maintenance of cerebral blood flow, normocapnia and the appropriate mean arterial pressure. In a patient with sickle cell disease, an adequate tissue perfusion, adequate oxygenation and hydration and strict pain control are to be primarily expected. The anesthesiologist is expected to know the physiopathology of both conditions to provide the best outcome for these patients.

PALABRAS CLAVE
Enfermedad de moya-moya;
Anemia;
Estenosis carotídea

Tratamiento anestésico en niño con enfermedad de moya-moya y anemia falciforme: caso clínico

Resumen
Objetivo: El objetivo de este caso clínico es el de describir el tratamiento anestésico en un niño con la enfermedad de moya-moya y anemia falciforme en un hospital terciario.

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http://dx.doi.org/10.1016/j.redar.2016.04.003
0034-9356/

Please cite this article in press as: Vinícius Ribeiro e Silva M, et al. Anesthetic management in a child with moya-moya disease and sickle cell anemia: Case report. Rev Esp Anestesiol Reanim. 2016. http://dx.doi.org/10.1016/j.redar.2016.04.003

Caso: Se trataba de un paciente de 6 años de edad, diagnosticado de enfermedad de moya-moya y anemia falciforme. Ambas condiciones se asocian a una elevada incidencia de episodios de isquemia cerebral. El paciente presentaba como antecedentes médicos 2 accidentes cerebrovasculares isquémicos. Se procedió a la administración de la anestesia para la realización de múltiples orificios craneales, y así favorecer la neovascularización en regiones cerebrales previamente hipoperfundidas.
Conclusión: Aunque escasamente descrito, el procedimiento anestésico aplicado en el paciente con enfermedad de moya-moya debe asegurar la estabilidad del flujo de sangre en el cerebro, normocapnia y el apropiado control de la presión arterial. En un paciente con anemia falciforme, la adecuada perfusión tisular, oxigenación e hidratación y el estricto control del dolor son los primeros objetivos a conseguir. El anestesiólogo debe conocer la fisiopatología de ambas condiciones para proveer el mejor cuidado de este tipo de pacientes.

Introduction

Although described in the whole world, moya-moya disease is a progressive cerebral vasculopathy that affects mainly the Asian population.[1] It has an yearly estimated incidence of 0.02–0.94 per 100,000 patients. It is characterized by the bilateral stenosis of the distal portion of the internal carotid artery or the proximal portion of the middle and anterior cerebral arteries.[1] The age distribution is bimodal, with one peak occurring in the first decade, and another in the second decade of life.[2] In children, it usually manifests itself through ischemic episodes, and in adults, through intracranial bleeding.[3]

Sickle cell anemia is a genetic hemoglobinopathy characterized by erythrocytes that, mainly under hypoxic conditions, assume a rigid sickle shape. It is more frequent in the black population. It is associated with a higher incidence of ischemic cerebrovascular accidents, particularly in the pediatric population.[4]

In this manuscript we report the case of a patient with moya-moya disease and sickle cell anemia, which, although they rarely occur together, deserve special attention from the anesthesiologist for the correct management during the perioperative period.

Case report

A female patient, 6 years old, black, 21 kg, diagnosed with sickle cell anemia since the neonatal period and with moya-moya disease, was electively scheduled for the execution of a neurosurgical procedure aiming the encephalic neovascularization in a tertiary hospital in Brasilia, Brazil. The legal guardian of the child reported an ischemic stroke episode at the age of 4 with the sequela of hemiparesis throughout the right hemibody, and a new ischemic stroke episode at the age of 5, with the patient remaining aphasic since then. Feeding is performed by means of a nasogastric tube. The child understands what is being said to her and obeys commands, but only expresses herself verbally through monosyllabic words. She denies having other systemic comorbidities, food or drug allergies. There is no history of sickle cell crisis and blood transfusion. She takes carbamazepine and clobazam. The magnetic resonance imaging of the skull revealed several bilateral ischemic lesions in the carotid area, with moya-moya type revascularization. The mother denies complications from sickle cell anemia. The physical examination revealed a child who was calm, acyanotic, afebrile, lucid and aware of time and space. Heart rate: 88; blood pressure: 98/50; Sp O2: 96% in room air; normal colored, hydrated, eupneic. There was no alterations on cardiopulmonary auscultation. Without predictors of difficult airway, with good oral opening and cervical mobility (Mallampati I). Additional exams revealed Hb: 12 g/dL, HTC: 35%, Na: 136 mmol/L, K^+: 3.8 mmol/L, Ca^{2+}: 0.9 mmol/L, Cl^-: 105.

In the operation room, venoclysis was performed in the right upper limb with a plastic catheter with a 18G needle after local infiltration anesthesia with lidocaine. An intravenous anesthesia was then performed with 70 μg of fentanyl, 25 mg of lidocaine without vasoconstrictor, 60 mg of propofol and 3 mg of cisatracurium. The tracheal intubation was performed with a number 3 curved Macintosh blade and a simple, number 5.5 endotracheal tube, without use of a guide. The cuff was inflated and the positioning was confirmed through capnography and symmetric bilateral auscultation. Arterial pressure: 95/48 mmHg. Attached to the ventilator under PCV, Pins 12 cmH_2O, respiratory rate: 15, PEEP: 4, FiO_2: 50%, realizing tidal volume Of 95 ml, maintaining a $EtCO_2$ of 33 mmHg. Anesthetic maintenance with an inspired fraction of 2% of sevoflurane. Subsequently, an urinary catheterization was performed to monitor the urinary output (residual urination of 25 ml). Hydration performed with SF 0.9%, totaling 250 ml and active warming by convection was provided.

The surgery consisted of a trichotomy in the bilateral parietal region, asepsis and antisepsis. The anesthesia was performed preemptively with bupivacaine 0.5% in the epidermis and dermis of the scalp. The incision and exposure of the skullcap was deperformed, where countless burr holes were made (Figs. 1 and 2).

At the end of the surgery, analgesia was performed with ketorolac 15 mg and dipyrone 1 g. Urination in the intraoperative period: 40 ml. The patient was decurarized with atropine 0.4 mg and neostigmine 0.8 mg, and extubated without complications. The entire surgical procedure

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