Rev Bras Anestesiol. 2016;xxx(xx):xxx–xxx

Sociedade Brasileira de Anestesiologia

REVISTA BRASILEIRA DE ANESTESIOLOGIA

Publicação Oficial da Sociedade Brasileira de Anestesiologia
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CLINICAL INFORMATION

Anesthetic management of a patient with 15q tetrasomy for dental treatment

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Received 10 June 2016; accepted 12 September 2016

KEYWORDS
Chromosome 15q tetrasomy;
General anesthesia;
Dental treatment

Abstract
Background and objectives: 15q tetrasomy is a chromosomal abnormality that is a part of the heterogeneous group of extra structurally abnormal chromosomes. This syndrome is characterized by epilepsy, central hypotonia, developmental delay and intellectual disability, and autistic behavior. This is the first report of the anesthetic management of a patient with this syndrome.
Case report: We administered general anesthesia for dental treatment in a patient with 15q tetrasomy.
Conclusions: Appropriate planning for the prevention of complications such as seizures and hypotonia, and for delayed emergence from anesthesia, is required. Specifically, choosing short-acting drugs that do not induce seizures, together with suitable monitoring, resulted in successful anesthetic management of the patient with 15q tetrasomy.

PALAVRAS-CHAVE
Tetrassomia do cromossomo 15q;
Anestesia geral;
Tratamento odontológico

Manejo anestésico de paciente com tetrassomia 15q para tratamento odontológico

Resumo
Justificativa e objetivos: Tetrassomia 15q é uma anomalia cromossômica que faz parte do grupo heterogêneo de cromossomos extras, estruturalmente anormais. Essa síndrome é caracterizada por epilepsia, hipotonia central, atraso no desenvolvimento e deficiência intelectual e comportamento autista. Este é o primeiro relato do manejo anestésico de um paciente com essa síndrome.
Relato de caso: Administramos anestesia geral para tratamento odontológico em um paciente com tetrassomia 15q.

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http://dx.doi.org/10.1016/j.bjane.2016.09.002

Please cite this article in press as: Hase Y, et al. Anesthetic management of a patient with 15q tetrasomy for dental treatment. Rev Bras Anestesiol. 2016. http://dx.doi.org/10.1016/j.bjane.2016.09.002

Conclusões: Um planejamento adequado para prevenir complicações como convulsões e hipotonia e para emergência tardia da anestesia é necessário. O manejo anestésico bem-sucedido do paciente com tetrassomia 15q foi o resultado especifico da escolha de medicamentos de curta duração que não induzem convulsões e monitorização adequada.

Introduction

Chromosome 15q tetrasomy is a constituent of the heterogeneous group of extra structurally abnormal chromosomes.[1] This syndrome displays distinctive clinical features, such as epilepsy, central hypotonia, developmental delay and intellectual disability, and autistic behavior.[1] Incidence at birth is estimated at 1 in 30,000; but no previous reports have described the anesthetic management of a patient with this syndrome.

Case report

The patient was a 25 year-old man (height, 153 cm; weight, 51 kg) who was scheduled for extraction of four impacted wisdom teeth and scaling under general anesthesia. He had a history of difficulty in sucking and feeding from birth and had started walking at the age of 2.5 years, because of developmental delay and hypotonia. He had no medical history of aspiration pneumonitis. He was diagnosed with 15q partial tetrasomy syndrome at the age of 8 years. He had experienced frequent generalized seizures since he was 7 years old, and had been treated with clobazam and carbamazepine. He showed autistic or autistic-like features. Although he had severe intellectual disability, he could speak one-word sentences and follow simple instructions. Blood examinations and ECG were unremarkable, except for slight liver dysfunction. He had a history of delayed emergence from general anesthesia 5 years earlier at our department.

On the day of the dental procedure, the patient entered the operating room and did not offer resistance to the monitors being worn. Slow induction was performed with 5% sevoflurane and 100% oxygen (6 L/min), following which an intravenous catheter was inserted and left in place, and fentanyl, thiamylal and rocuronium were administered. After uneventful mask ventilation, nasal intubation was performed with a 6.5 mm cuffed Nasal RAE™ endotracheal tube under direct visualization of the glottis. General anesthesia was maintained with air, oxygen, desflurane (fraction of inspired oxygen = 0.4) and remifentanil under controlled ventilation. Immediately after the induction of anesthesia, the Train Of Four Ratio (TOFR) was 0, increasing to 0.6 after 10 min. By the start of the surgery, it had reached 0.9. The Bispectral Index (BIS) value during the surgery was maintained between 50 and 65, without any unexpected large variations in the value. His vital signs remained stable with a blood pressure of 80–90/40–50 mmHg, heart rate of 60 bpm, SpO_2 of 99%–100%. Sugammadex was administered after the surgery and extubation was performed after confirming that muscle function was fully restored, as indicated by a TOFR of 1.0. The operative time was 74 min and anesthesia time was 146 min. The patient was discharged on the following day without any problems such as perioperative respiratory complications and seizures.

Discussion

Chromosome 15q tetrasomy is also known as inverted duplication 15 (inv dup[15]) or isodicentric chromosome 15 (idic[15]).[2] It is caused by an extra inv dup(15) marker chromosome, cytogenetically defined as inv dup(15) (pter → q12-13: q12-13 → pter) by FISH analysis.[1] Our patient's karyotype was 47, XY, + idic(15)(q12), i.e. tetrasomy with the extra derived chromosome at 15q11-q12. 15q tetrasomy is clinically associated with seizures, hypotonia, developmental delays, intellectual disability, congenital heart defects, brain abnormalities, and dysmorphic facial features, such as downslanting palpebral fissures, low set ears, micrognathia, prominent mandible, and cleft palate.[1,2] 15q11-q13 is considered a prime candidate region for the mapping of epilepsy genes and the relationship with autism.[2,3] Gene tetrasomy may alter GABA receptor activity upon which the major CNS inhibitory mechanisms rely.[1] 15q tetrasomy affects encoding of $GABA_A$ receptor subunits (GABRB3, GABRA5, GABRG3), which is related to the seizures and autism associated with this syndrome.[3] Some reports show high complication rates of seizures in 15q tetrasomy patients (63%–75%).[1,3] Therefore, the challenges for anesthesiologists managing such patients are attacks of seizures, perioperative respiratory complication and prolonged muscle relaxation related to hypotonia, and difficult airways.

The following five points were considered when we planned the anesthetic management for the present patient. First, for the prevention of seizures, the patient took his regular antiepileptic medicines on the day of surgery, and thiamylal, which has anticonvulsant effects, was injected at the induction of anesthesia. We also had to examine the medicines which were substituted for the regular ones in case the patient could not ingest orally perioperatively. Desflurane is a suitable drug for use in patients with epilepsy since it does not have epileptiform activity[4] while there have been several reports of epileptoid EEG patterns observed under sevoflurane anesthesia.[5] It is quite difficult to observe a seizure during general anesthesia. However, previous reports have shown that abnormal fluctuations or decreases in BIS values under anesthesia resulted from occurrences of epileptiform EEG activity, and that BIS monitoring might not only give useful information on the sedative-hypnotic state,

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