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CLINICAL INFORMATION

Atypical reaction to anesthesia in Duchenne/Becker muscular dystrophy

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KEYWORDS
Duchenne muscular dystrophy;
Malignant hyperthermia;
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Abstract
Background and objectives: Duchenne/Becker muscular dystrophy affects skeletal muscles and leads to progressive muscle weakness and risk of atypical anesthetic reactions following exposure to succinylcholine or halogenated agents. The aim of this report is to describe the investigation and diagnosis of a patient with Becker muscular dystrophy and review the care required in anesthesia.
Case report: Male patient, 14 years old, referred for hyperCKemia (chronic increase of serum creatine kinase levels – CK), with CK values of 7779–29,040 IU L^{-1} (normal 174 IU L^{-1}). He presented with a discrete delay in motor milestones acquisition (sitting at 9 months, walking at 18 months). He had a history of liver transplantation. In the neurological examination, the patient showed difficulty in walking on one's heels, myopathic sign (hands supported on the thighs to stand), high arched palate, calf hypertrophy, winged scapulae, global muscle hypotonia and arreflexia. Spirometry showed mild restrictive respiratory insufficiency (forced vital capacity: 77% of predicted). The in vitro muscle contracture test in response to halothane and caffeine was normal. Muscular dystrophy analysis by Western blot showed reduced dystrophin (20% of normal) for both antibodies (C and N-terminal), allowing the diagnosis of Becker muscular dystrophy.

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Conclusion: On preanesthetic assessment, the history of delayed motor development, as well as clinical and/or laboratory signs of myopathy, should encourage neurological evaluation, aiming at diagnosing subclinical myopathies and planning the necessary care to prevent anesthetic complications. Duchenne/Becker muscular dystrophy, although it does not increase susceptibility to MH, may lead to atypical fatal reactions in anesthesia.

PALAVRAS-CHAVE
Distrofia muscular de Duchenne;
Hipertermia maligna;
Anestesia

Reação atípica à anestesia em distrofia muscular de Duchenne/Becker

Resumo
Justificativa/objetivos: Distrofia muscular de Duchenne/Becker afeta a musculatura esquelética e leva a fraqueza muscular progressiva e risco de reações atípicas anestésicas após exposição à succinilcolina ou halogenados. O objetivo do presente relato é descrever investigação e diagnóstico de paciente com distrofia muscular de Becker e revisar os cuidados necessários na anestesia.
Relato de caso: Paciente masculino, 14 anos, encaminhado por hiperCKemia (aumento crônico dos níveis séricos de creatinoquinase – CK), com valores de CK de 7.779–29.040 UI.L^{-1} (normal 174 UI.L^{-1}). Apresentou discreto atraso da aquisição de marcos motores (sentou aos nove meses, andou aos 18). Antecedente de transplante hepático. No exame neurológico apresentava dificuldade para andar nos calcanhares, levantar miopático (apoiava mãos nas coxas para ficar de pé), palato arqueado alto, hipertrofia de panturrilhas, escápulas aladas, hipotonia muscular global e arreflexia. Havia insuficiência respiratória restritiva leve na espirometria (capacidade vital forçada: 77% do previsto). O teste de contratura muscular *in vitro* em resposta ao halotano e à cafeína foi normal. Estudo da distrofina muscular por técnica de Western *blot* mostrou redução da distrofina (20% do normal) para ambos os anticorpos (C e N-terminal), e permitiu o diagnóstico de distrofia muscular de Becker.
Conclusão: Na avaliação pré-anestésica, história de atraso do desenvolvimento motor, bem como sinais clínicos e/ou laboratoriais de miopatia, deve motivar avaliação neurológica, com o objetivo de diagnosticar miopatias subclínicas e planejar cuidados necessários para prevenir complicações anestésicas. Distrofia muscular de Duchenne/Becker, apesar de não conferir suscetibilidade aumentada à HM, pode levar a reações atípicas fatais na anestesia.

Introduction

The dystrophin protein stabilizes sarcolemma in the skeletal, cardiac and smooth muscle, and central nervous system; therefore its absence/decrease alters the sarcolemma structure, allows Ca^{2+} influx, intracellular protease activation, and muscle fiber necrosis.[1,2] Duchenne muscular dystrophy is a myopathy that affects one in 3600 live births as a result of mutations in the dystrophin gene, which leads to its absence with a recessive inheritance linked to the X chromosome.[1] In Becker muscular dystrophy, mutations in the dystrophin gene allows expression of the protein, although abnormal.[1] Patients with Duchenne/Becker muscular dystrophy present with progressive necrosis of skeletal muscle that begins in childhood so the diagnosis may go unnoticed in the first years of life.[1]

In these patients, exposure to succinylcholine and halogenated agents may be followed by atypical reactions in anesthesia and even sudden cardiac arrest due to hyperkalemia resulting from massive rhabdomyolysis.[1]

Objective

The aim of this report is to describe the investigation and diagnosis of a patient with Becker muscular dystrophy and review the anesthetic care needed.

Case report

A 14-year-old male patient was referred to the malignant hyperthermia service for hyperCKemia (a chronic increase of serum creatine kinase levels – CK) investigation, with CK values in the three years prior to the consultation ranging from 7779 to 29,040 UI L^{-1} (normal value 174 IU L^{-1}).

After an uneventful gestation, the patient was delivered via cesarean section due to dystocia, with report of transient neonatal jaundice. He presented a mild delay in motor milestones (sitting at 9 months and walking at 18 months). At the age of three months, jaundice, dyslipidemia and coagulopathy were detected, what allowed

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