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

Sociedade Brasileira de Anestesiologia

REVISTA BRASILEIRA DE ANESTESIOLOGIA

Publicação Oficial da Sociedade Brasileira de Anestesiologia
www.sba.com.br

CLINICAL INFORMATION

# Anesthetic management of two patients with alkaptonuric ochronosis for total knee arthroplasty

**Betul Kozanhan**

*Konya Education and Research Hospital, Department of Anesthesiology and Reanimation, Konya, Turkey*

Received 2 February 2016; accepted 27 July 2016

**KEYWORDS**
Alkaptonuria;
Ochronosis;
Anesthesia

**Abstract** The current case report describes two cases of alkaptonuric ochronosis for anesthetic management. Alkaptonuria is a rare genetic orphan disease of tyrosine metabolism characterized by an accumulation of homogentisic acid in cartilage and connective tissues. Patients present most commonly for orthopedic joint surgery due to progressive arthropathy that can be misdiagnosed many a times. However respiratory, airway, cardiovascular and genitourinary systems' complications can occur with age progressing. Restricted range of motion of cervical spine may lead to difficulty with airway management. In addition, degenerative changes and stiffness of lumbar spine due to ochronosis would make neuraxial blockade challenging. Although this inherited condition is extremely rare, anesthesiologists should be aware of its existence and prepare for management of potential challenging problems. This report highlights special care and precautions that need to be taken during anesthetic management.
© 2016 Sociedade Brasileira de Anestesiologia. Published by Elsevier Editora Ltda. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

**PALAVRAS-CHAVE**
Alcaptonúria;
Ocronose;
Anestesia

**Tratamento anestésico de dois pacientes com ocronose alcaptonúrica para artroplastia total do joelho**

**Resumo** Este relato descreve o tratamento anestésico em dois casos de ocronose alcaptonúrica. Alcaptonúria é uma doença genética rara do metabolismo de tirosina caracterizada por acúmulo de ácido homogentísico em cartilagem e tecidos conjuntivos. Os pacientes geralmente recorrem à cirurgia ortopédica devido à artropatia progressiva que, muitas vezes, pode ser diagnosticada incorretamente. No entanto, complicações das vias respiratórias, cardiovasculares e geniturinárias podem ocorrer com o avanço da idade. A restrição de mobilidade da

*E-mail:* betulkozanhan@gmail.com

http://dx.doi.org/10.1016/j.bjane.2016.07.015
0104-0014/© 2016 Sociedade Brasileira de Anestesiologia. Published by Elsevier Editora Ltda. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Please cite this article in press as: Kozanhan B. Anesthetic management of two patients with alkaptonuric ochronosis for total knee arthroplasty. Rev Bras Anestesiol. 2016. http://dx.doi.org/10.1016/j.bjane.2016.07.015

coluna cervical pode levar ao manejo difícil das vias aéreas. Além disso, as alterações degenerativas e a rigidez da coluna lombar devido à ocronose podem tornar o bloqueio neuroaxial um desafio. Embora essa condição hereditária seja extremamente rara, os anestesiologistas devem estar cientes de sua existência e se prepararem para o manejo de potenciais problemas desafiadores. Este relato destaca os tratamentos e precauções especiais que devem ser tomadas durante o manejo anestésico.
© 2016 Sociedade Brasileira de Anestesiologia. Publicado por Elsevier Editora Ltda. Este é um artigo Open Access sob uma licença CC BY-NC-ND (http://creativecommons.org/licenses/by-nc-nd/4.0/).

## Introduction

Alkaptonuria (AKU) is a rare metabolic disorder, with an estimated prevalence between 1 in 250,000 and 1 million live births in most populations.[1] AKU is caused by a deficiency of the Homogentisate 1,2-dioxygenase (HGO) enzyme, which converts homogentisic acid (HGA) to maleylacetoacetic acid in the catabolic pathway of tyrosine. Disease is characterized by the accumulation of HGA and its oxidized products in all connective tissues of the body. Accumulation of HGA may act as a chemical irritant, leading to inflammation and degeneration. As a result, AKU has three major features; (1) homogentisic aciduria (darkening of the urine upon contact with air or after exposure to an alkaline agent); (2) ochronosis (bluish-black pigmentation of connective tissue); and (3) arthritis.[1]

Ochronotic arthropathy is the most common complication of AKU.[1] The deposition of polymer is assumed to cause an inflammatory response that results in calcium deposition in both synovial and intervertebral joints. Clinical manifestations include back and peripheral joint pain, limitation of movement and stiffness. Joint symptoms, typically begin in the large weightbearing joints at third decade of life, and progress, until chronic pain prompts a knee, hip, or shoulder replacement, at an average age of 55 years.[1] Valvular heart diseases,[1,2] renal stones,[1] restrictive pulmonary disease are other important but less common consequences of alkaptonuric ochronosis.[1] There is no definitive cure for AKU and treatment management is usually symptomatic. In this case report we describe the anesthetic management of two patients with AKU for a total knee replacement surgery.

## Case 1

A 59-year-old, body mass index (BMI) = 44.6 kg/m$^2$ female patient complained of low back pain with stiffness, bilateral knee pain causing restricted mobility. She was diagnosed with end-stage knee arthritis and scheduled for left knee replacement surgery. A review of her medical history revealed hypertension and diabetes mellitus. She had previously undergone bilateral hip replacement surgery (at ages 49 and 48). Preoperative airway examination showed adequate mouth opening, Mallampati classification MP-III, 5.5 cm of thyromental distance, short, thick neck, with limited cervical spine mobility. The cardiac and respiratory systems were normal on examination. Complete blood count, coagulation profile, biochemical parameters, renal and liver function were also within normal limits. She was American Society of Anesthesiologists (ASA) physical class III. We initially planned to perform surgery under spinal anesthesia and written informed consent was obtained. In the operating room standard monitors (ECG, pulse oximeter, non-invasive blood pressure) were applied and an intravenous infusion of normal salin solution started at a rate of 15 mL/min. During positioning we faced with difficulty due to her multiple joint deformities. After premedication (midazolam 2 mg and fentanyl 50 mcg) and taking all aseptic precautions lumbar punctures were attempted at 2 different spinal levels (L3–4 and L4–5) using by median or paramedian approach by two experienced anesthesiologists. But unfortunately flexibility of the lumbar spine was reduced and our attempts failed as the needle was met with bone in all directions. The technique of awake intubation under local anesthesia was explained to the patient and consent was obtained. Our initial plan was to use fiberoptic methods, but the equipment was unavailable, so the videolaryngoscope was used instead. The patient oropharyngeal mucosa was anesthetized by 10% lidocaine topical spray; fentanyl 50 μg and atropine 0.5 mg were administered intravenously. The tracheal intubation was performed at the first attempt using the McGrath videolaryngoscope without any complications. A Grade 3 view (Cormack and Lehane) of the larynx was obtained and 7.0 cuffed endotracheal tube was passed through the larynx over difficult intubation stylet. The surgery lasted 55 min, patient remained hemodynamically stable throughout the procedure. There was minimal blood loss, approximately 1700 mL of normal salin was infused. During surgery, black discoloration was visible on the joint cartilage and a concern was raised that this patient may have ochronosis. The patient was extubated uneventfully upon full awakening from anesthesia and transported to the recovery room. She was discharged home on the third postoperative day without any complications. After the histological diagnosis, the patient was re-examined for alkaptonuria. She reported a history of dark brown discolored urine but she has never complained about it before. There were black ochronotic pigmentations of the sclera and skin.

## Case 2

A 57-year-old, BMI = 24.4 kg/m$^2$ male patient, scheduled for left knee replacement surgery with a diagnosis of alkaptonuric ochronosis. A review of his medical history revealed

Please cite this article in press as: Kozanhan B. Anesthetic management of two patients with alkaptonuric ochronosis for total knee arthroplasty. Rev Bras Anestesiol. 2016. http://dx.doi.org/10.1016/j.bjane.2016.07.015

Download English Version:

https://daneshyari.com/en/article/8611654

Download Persian Version:

https://daneshyari.com/article/8611654

Daneshyari.com