CASE REPORTS

THE USE OF NEAR INFRARED SPECTROSCOPY IN ALKAPTONURIA - THE MISLEADING OCHRONOSIS - A CASE REPORT AND LITERATURE REVIEW

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Abstract

Introduction: *Near infrared spectroscopy is a non-invasive method to assess regional oxygenation and is being used in transcatheter aortic valve implantation to document periods of cerebral hypoperfusion, where cerebrovascular events are one of the most feared complications.*

Alkaptonuria is a rare metabolic disease characterized by accumulation of homogentisic acid in tissues and body fluids. The accumulation of pigment might interfere with the absorption of near infrared light, used in near infrared spectroscopy monitoring.

We present a case of near infrared spectroscopy failing to accurately monitor cerebral oximetry in a woman, with alkaptonuria, undergoing a transcatheter aortic valve implantation.

INTRODUCTION

Near infrared spectroscopy (NIRS) is a non-invasive method to assess cerebral regional oxygen saturation (rSO2) and has been widely used in perioperative management of patients submitted to cardiac procedures1. Aside from its use in cardiac surgery, NIRS technology is now used in percutaneous procedures, such as transcatheter aortic valve implantation (TAVI). TAVI is an alternative treatment in inoperable, high or intermediate risk patients with aortic stenosis[2].

One of the most feared complications of TAVI is the occurrence of cerebrovascular (CV) events in perioperative setting. Stroke rates have declined over the years, due to improvements in valve technology and surgical technique, and current rates range from 1 to 11% within 30 days. Despite of that, stroke is associated with increased morbidity and mortality[3]. Furthermore, a proper hemodynamic management is essential for the success of the procedure and to decrease complications. For these reasons, the use of cerebral rSO2 can be used to guide hemodynamic management and to provide evidence of cerebral hypoperfusion[4]. However, one of the limitations of using such monitors is the interference of extracranial tissue with the reading of the signal[5].

Alkaptonuria is a rare metabolic disease with a prevalence of 1 case in 250 000–1 000 000 births, with a recessive inheritance. It results from the absence of homogentisate 1,2 dioxygenase, the enzyme responsible for the breakdown of homogentisic acid (HA). The accumulation of HA in tissues and body fluids results in typically dark urine and ochronosis - darkening of collagenous tissues. The main complications are ochronotic osteoarthropathy, cardiac valve disfunction, chronic kidney disease, calculi and characteristic discoloration of skin[6]. We present a case about the inability to accurately monitor cerebral rSO2 in a patient with alkaptonuria who underwent TAVI.

CASE DESCRIPTION

A 78 year-old female, ASA III, was scheduled to TAVI due to severe aortic stenosis. She had a history of type 2 diabetes mellitus and alkaptonuria with target organ disfunction: chronic kidney disease, renal calculi, ochronotic arthrop-

athy and severe aortic stenosis. The standard preoperative workup showed a severe calcified stenotic aortic valve with a valve area of 0,4cm2, a peak and medium transvalvular pressure gradient of 114/78mmHg, respectively, with a moderate depression of left ventricular function documented on transesophageal echocardiography, and no associated coronary artery disease. The preoperative laboratory values were unremarkable apart from a low hemoglobin value (11,6 g/dL) and a glomerular filtration rate of 61mL/min/1.73m2. As for previous surgical interventions there was only a cataract surgery with local anesthesia. Physical examination revealed a dark brown pigmentation of the face and eyes (figure 1).

On arrival to the hemodynamic laboratory, the patient was monitored according to ASA standards. Cerebral oximetry was monitored using bilateral pads (INVOS® 5100C, Medtronic). Standard cleansing measures were taken before applying the pads. Pads were placed over the frontal area, bilaterally, and the baseline value was recorded. The baseline value was 15, bilaterally and the value remained unchanged after supplementation of oxygen by a nasal cannula (figure 2). The whole INVOS® system was changed (pads, cables and monitor) but the value remained the same. A 6-Fr introducer was inserted in the right internal jugular vein to pass the pacemaker wires. The TAVI procedure was performed under conscious sedation, using a target-controlled infusion of propofol and remifentanil. Femoral vascular access was used and the implantation of an Edwards Sapiens 23mm valve was uneventful. Throughout the procedure the INVOS value remained unchanged. No adverse events were recorded. After the procedure, the patient went to the intensive care unit, awake and with a normal neurological exam.

DISCUSSION

TAVI is associated with the risk of cerebrovascular events and the use of NIRS monitoring has been used to detect cerebral hypoperfusion. NIR light spectrum is used for monitoring because it has the ability to pass biological tissues and to be absorbed by chromophores, hemoglobin being the most important. However, the presence of other chromophores can interfere with the light transmission. Although the amount of NIR light absorbed correlates with chromophore concentration, we assumed, in this case, that NIR light wasn't able to penetrate patient's forehead and reach the cortex.

Argiriadou et al reported one similar case and assumed that the pigmentation of skin and the accumulation of pigment in the periosteum or dura mater might explain the impossibility of NIR light to penetrate tissues. However, we still don't know the absorption spectrum of HA, neither if it has significant absorption at the wavelengths used by INVOS® device.

There are several NIRS devices available that differ in the algorithms used, the wavelengths of light emitted and the number and distance between the light emitters and detectors[8]. The INVOS® device uses NIR light at 2 wavelengths, has one emitting diode and two detectors. On the other hand, EQUANOX® has two emitting diodes, two detectors and uses 4 wavelengths of NIR. For this reason, is able to more accurately eliminate extracranial contamination of the signal, being less affected by concentration of other chromophore or surface defects.

There are few cases reported about the inability to accurately monitor rSO2 in patients with alkaptonuria[9-10]. Regarding the devices used in all studies, only EQUANOX® might be used in alkaptonuria, since it's less affected by extracranial contamination.

Alkaptonuria is strongly associated with valvular disease requiring surgical procedures. These high-risk patients submitted to cardiac procedures benefit from cerebral oximetry monitoring. More research should be taken to improve the accuracy of this monitors, to enable its use in patients with alkaptonuria.

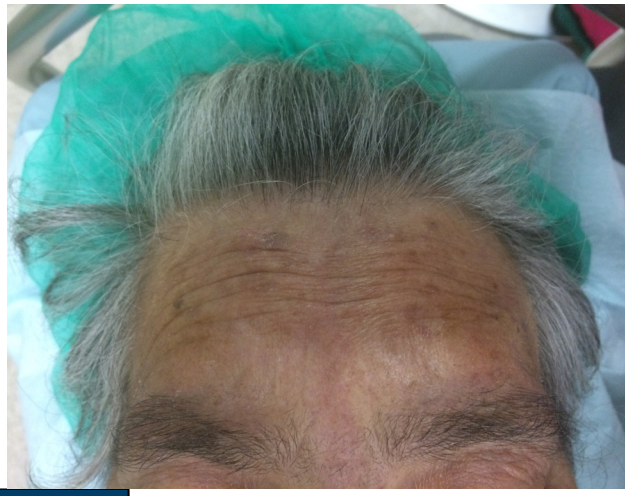

Figure 1 *Dark pigmentation of patient's forehead*

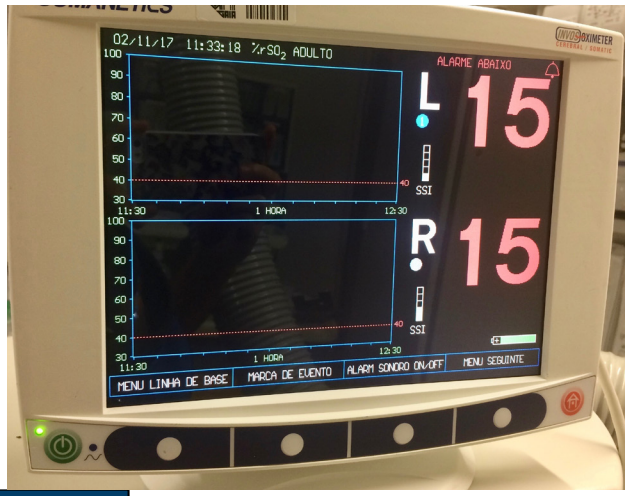

Figure 2 *INVOS monitor; Baseline value.*

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