Anaesthetic management of a Greek patient with congenital insensitivity to pain and anhidrosis undergoing a scheduled orthopaedic operation

Paraskevi Matsota, Ageliki Pandazi, Marilia Loizou, Georgia Kostopanagiotou

2nd Department of Anaesthesiology, School of Medicine, University of Athens, “Attikon” Hospital, Athens, Greece

Submitted: 11 May 2008
Accepted: 14 August 2008

Arch Med Sci 2009; 5, 1: 115-116
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Abstract

Congenital insensitivity to pain with anhidrosis (CIPA) is a very rare hereditary syndrome worldwide due to certain gene mutations and histological appearances that affect the peripheral pathway of noxious stimuli transmission and the innervation of sweat glands. It is characterized by insensitivity to pain, anhidrosis, and heat intolerance. We describe the clinical features and the anaesthetic management of a 24-year-old Greek man suffering from CIPA, who uneventfully underwent an internal osteosynthesis of the femoral bone under general anaesthesia. This is also the first reported case with congenital insensitivity to pain and anhidrosis in Greece.

Key words: anaesthesia, congenital pain insensitivity and anhidrosis.

Introduction

Congenital insensitivity to pain with anhidrosis (CIPA) is a rare autosomal recessive disorder caused by mutations in the neurotrophic tyrosine receptor kinase 1 gene, which is the receptor for nerve growth factor [1]. It is characterized by the clinical triad of insensitivity to pain, anhidrosis, and heat intolerance. Pathological findings include loss of small-calibre (A-delta and C) nociceptive nerve fibres, lack of small neurons in the dorsal root ganglia, absence of the Lissauer tract and loss of innervation of the sweat glands.

In contrast to the former belief [2-4] that CIPA patients could safely be managed with anaesthesia, enhanced risk of cardiovascular events has been reported [5]. We describe the anaesthetic management of a Greek patient with CIPA.

Case report

A 24-year-old Greek male with established CIPA, ASA physical status II, weighing 62 kg, was scheduled for an internal osteosynthesis for femoral bone fracture. He was diagnosed in infancy after he cut himself with a piece of glass and experienced no pain. Histological studies at that time confirmed CIPA. He also could not discriminate heat from cold, but he could sense itching, titillation and he had tactile hyperesthesia mainly localized...
around both patellae. He also reported tension-type headaches mainly associated with anxiety, which could be treated with paracetamol.

Metoclopramide 10 mg and ondansetron 4 mg were administered before anaesthesia induction. General anaesthesia was induced with midazolam 2 mg, propofol 200 mg, and rocuronium bromide 60 mg, and was maintained with sevoflurane in air/oxygen mixture. Intraoperative monitoring included electrocardiogram, heart rate, non-invasive blood pressure, SpO₂, end-tidal CO₂, oesophageal temperature monitoring, peripheral nerve stimulator, and BIS monitoring (A-2000 monitor Aspect Medical systems, Natick, MA, USA). A heating/cooling blanket was used to maintain the patient’s body temperature. Surgery lasted for 3 h. Intraoperatively, 1.6 l of Ringer’s lactate and 500 ml of hydroxyethyl starch solution (6% HES 130 000/0.4) were infused to restore volume deficit due to h preoperative fasting (640 ml), blood loss (600 ml), and the hourly requirements, while the patient retained stable (BP 104/50 mm Hg, HR 55-60 bpm, SpO₂ 99-100%, ETCO₂ 28-31 mm Hg, temperature 36.1°C). BIS value was in the range 35-54 with sevoflurane concentrations ranging from 1.2 to 1.5 MAC. Tracheal extubation was achieved while the patient retained stable (BP 104/50 mm Hg, HR 55-60 bpm, SpO₂ 99-100%, ETCO₂ 28-31 mm Hg, temperature 36.1°C). Bispectral index (BIS) values were maintained at 40-80.

In patients with CIPA, strict perioperative temperature control is the primary anaesthetic goal, while administration of anticholinergic drugs is not recommended due to the risk of hyperthermia. Consequently we avoided anticholinergics. Postoperative nausea and vomiting (PONV) has also been reported in CIPA patients [2]. For our patient, the pre-anaesthetic administration of metoclopramide and ondansetron may have played a role in preventing PONV. In some clinical cases, it was also expressed that these patients often have cardiovascular complications when following anaesthesia, despite the kind of used anaesthetic drugs or the type of surgical procedure [5]. In this retrospective study from Israel, a high incidence of intraoperative hypotension and bradycardia including an intraoperative death was reported. In CIPA patients, the possibility of autonomic system dysfunction must be taken into consideration, while decreased norepinephrine and dopamine levels have been reported in these patients [7]. In our case, the absence of cardiovascular complications intraoperatively could be the result of the appropriate volume replacement and the titration of the anaesthetics given. However, our patient did not mention disturbance of gastrointestinal motility, or episodes of bradycardia or tachycardia; thus lack of autonomic dysfunction should be taken into account.

In conclusion, our suggestions for the best approach to CIPA patients include: assessment of autonomous dysfunction, establishment of haemodynamic stability and normothermia, and elimination of PONV and awareness.

References