Anesthetic management of a pediatric patient with hypohidrotic ectodermal dysplasia undergoing emergency surgery

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Received 16 October 2013; accepted 31 October 2013
Available online 11 December 2013

Abstract  Ectodermal dysplasias are rare conditions with a triad of hypotrichosis, anodontia and anhidrosis. In literature review there have been only a few reports of anesthetic management of patients with ectodermal dysplasias. Hyperthermia is a very serious risk which may occur due to the defect of sweat glands. The present case involves a 10-year-old child with ectodermal dysplasia who presented with an acute abdomen and was considered for an emergency surgery. Our aim was to demonstrate the successful management of this case using a combination of general and epidural anesthesia. It is important for anesthesiologist to have information about this syndrome in case of emergency operations, since it can prevent serious complications and even save lives.

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Palavras-chave
Displasia ectodérmica; Anestesia regional; Abdome agudo

Manejo anestésico de paciente pediátrico com displasia ectodérmica hipoidrótica submetido a cirurgia de emergência

Resumo As displasias ectodérmicas são condições raras, com uma triade de hipotricose, anodontia e anidrose. Em revisão da literatura há apenas alguns relatos de manejo anestésico de pacientes com displasias ectodérmicas. Hipertermia é um risco muito sério que pode ocorrer por causa de defeito das glândulas sudoríparas. O presente caso envolve uma criança de 10 anos com displasia ectodérmica que se apresentou com abdome agudo e foi considerada para uma cirurgia de emergência. Nosso objetivo foi demonstrar o manejo bem-sucedido desse caso, com o uso de uma combinação de anestesia geral e peridural. É importante para o anestesiologista

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Introduction

Hypohidrotic ectodermal dysplasia (HED) (Christ-Siemens-Touraine syndrome) is an X-linked condition and is the most common form of ectodermal dysplasia (ED). In this type of syndrome, patients have no sweat glands or they are significantly decreased. HED has characteristic triad of reduction in the amount of hair (hypotrichosis), absence of sweat glands (anhidrosis) and hypodontia.1

Patients with HED frequently suffer from pulmonary infections and hyperthermia in increased ambient temperatures.2

Case report

A 10-year-old boy with HED presented on our pediatric surgery emergency department with a 12-h history of nausea and vomiting associated with right lower quadrant abdominal pain. Abdominal examination revealed right lower quadrant tenderness with guarding and rebound. Child has characteristic triad of HED (Figs. 1 and 2). Laboratory testing revealed a mild leukocytosis of 13,400 cells/mm³. All other laboratory results showed normal values. Patient’s past medical history was significant for HED. Patient had no surgical history. The patient was diagnosed with appendicitis and emergency surgery was planned.

Premedication included intravenous midazolam (0.15 mg/kg). The patient was transported to the operating room and routine monitors were placed. Preanesthetic airway examination revealed Mallampati class-I in sitting and supine position. Although we didn’t suspect difficult airway we prepared difficult airway cart which included different sizes of endotracheal tubes, laryngeal mask airways, proeseal LMA and fiberoptic bronchoscope.

After preoxygenation with 100% oxygen, anesthesia was induced with propofol (3 mg/kg) and fentanyl (2 μg/kg). After ventilation, neuromuscular blockade was provided by rocuronium (1 mg/kg). Laryngeal view with the Macintosh laryngoscope was Cormack–Lehane grade-I and the patient’s trachea was intubated easily. At the beginning of operation body temperature was 36.0 °C. His body temperature remained 36–36.5 °C during the operation. After intubation the patient was positioned in the left lateral position and the skin was prepared with polivinilpiloridon iot. An 18 G epidural catheter was sited without difficulty at the level of the third and fourth lumbar interspace using the loss of resistance technique a test dose of levobupivacaine 5 mg was administered. 10 mL of levobupivacaine 0.125% was infused into the epidural space. Anesthesia was maintained with propofol and epidural

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analgesia. The body temperature monitored continuously. Repeated dosing of the neuromuscular blockade was not necessary.

Appendectomy was performed in normal course without an incident within 45 min. Following completion of the surgical procedure, residual neuromuscular blockade was reversed with sugammadex 100 mg and the patient’s trachea extubated. Patient was discharged from the hospital two days after the operation.

Discussion

HED is the most frequent form of ED and a rare syndrome with an incidence of approximately 1:100,000. Hyperthermia due to inadequate sweating, secondary to partial or complete absence of sweat glands, is common in the anhidrotic form and results in a potential for sudden death in infancy due to hyperthermia. The reported mortality rate is in the order of 10–54%.

Problems for anesthesia in a person with the HED include potentially difficult tracheal intubation due to multiple missing teeth and hypoplasia of the maxilla. Respiratory function needs to be assessed because of recurrent chest infections as well.

In PubMed databases literature review there have been only a few reports of anesthetic management of patients with HED.

Hotta et al. emphasized airway hydration, preparation for difficult airway and following up body temperature in a case report which they mentioned management of anesthesia for ophthalmic surgery of a 10-year-old child that have HED. Also Sugi et al. reported a case which they used epidural anesthesia for skin grafts and debridement in a 20-year-old male patient with HED. Docquier et al. add lumbar epidural analgesia to protocol used intravenous anesthesia in an elective lower extremity orthopedic surgery case which was applied to an 8-year-old girl with multiminicore myopathy, andED. They reported this combination provided a stable anesthesia, without any complications and constituted an excellent analgesia during operation and post-operative period.

Until today all reported cases were about scheduled elective surgeries and therefore they had time to prepare. In our case, patient was diagnosed by a pediatric surgeon with an acute abdomen in ER and therefore was a surgical emergency situation and the patient was diagnosed with HED during the preanesthetic visit just before the surgery. So, there was not enough time to research or get more information on HED. Surgical procedure was completed successfully without any complications via epidural analgesia combined with intravenous anesthesia. We believe that regional anesthesia techniques can be quite useful in these cases. It is important to pay attention for airway humidification and difficult intubation. In the operation theater, difficult airway cart must prepared. No halogenated inhalation anesthetics and depolarizing muscle relaxants were used. Body temperature was monitored throughout the case.

In conclusion, HED is a challenge for anesthesiologists and critical care physicians during the perioperative time. The risk of malignant hyperthermia to anesthetic drugs should be kept in mind. All anesthesiologists must be knowledgeable in the case of having no time for preparation and making quick decisions for the most appropriate anesthesia method in emergency surgical interventions such as trauma and acute abdomen.

Conflicts of interest

The authors declare no conflicts of interest.

References