

Anesthesia in a pediatric patient with Xeroderma pigmentosoma: A case report

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Abstract

Xeroderma pigmentosum (XP) is an autosomal recessive genetic disorder with the following conditions: neurologic disorders that gradually progress and skin hypersensitivity to ultraviolet (UV). Inhalational anesthetic drugs should be avoided in these patients since they may induce DNA damage, also use of muscle relaxants should be kept to a minimum. Thus for XP patients, total intravenous anesthesia (TIVA) is more appropriate for inducing general anesthesia and airway manipulation must be kept to a minimum. We report a 6 year old boy with XP and its airway management and anesthesia during surgery.

Keywords

- Xeroderma pigmentosum
- general anesthesia
- total intravenous anesthesia

Introduction

Xeroderma pigmentosum (XP) is an autosomal recessive genetic disorder¹ with the following conditions: neurologic disorders that gradually progress, ocular involvement and skin hypersensitivity to sunlight.²

The incidence varies considerably in different parts of the world, the lowest being in Western Europe and the highest being in North Africa and the Middle East.³ It is believed that volatile anesthetics may induce DNA damage in the cells of patients with XP and aggravate their symptoms,⁴ although no human data exist regarding this effect up to

now.⁵ Minimum muscle relaxation is used during the operation. Patients with XP may have difficult airway.⁶

Case Presentation

A 6-year-old boy with XP was referred to our institute for excisional biopsies of several malignant skin tumors. Since he was three years old he had extensive brown skin lesions and tumors. The tumors near the orbit and the lower eyelid required excisional biopsy and were replaced by skin grafts from the medial region of the right arm. In primary evaluation, he was completely alert and oriented. He had no prior medical history of neurological

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signs and symptoms seizures. When evaluating the airway, mouth opening was limited also he had a grade II Mallampati but there were no mucosal lesions.

Premedication was performed using IV midazolam and Fentanyl. Monitoring was carried out using pulse oximetry, ECG and blood pressure. Pre-oxygenated was performed with 100% oxygen for about 3 minutes. Induction was carried out using lidocaine and propofol. LMA size 2 was used for intubation and proper placement was confirmed by auscultation over the neck.⁷ Maintenance of anesthesia was achieved with N₂O 50%, O₂ 50% and propofolremifentanil infusion. He did not receive any muscle relaxants and had spontaneous ventilation. He did not experience any adverse effect during the operation or recovery **Figure 1**.



Figure 1: Patient under anesthesia

Discussion

Xeroderma pigmentosum is an autosomal recessive genetic disorder with its unique characteristics being: neurological disorders, very high risk of skin cancer and hypersensitivity to sunlight.⁸ This disorder was originally described in 1870

by Kaposi; it presents itself in childhood and has premalignant and malignant lesions which eventually lead to death in adulthood. These patients have many types of abnormalities such as facial and oropharyngeal defects which lead to difficult intubation, prolongation of neuromuscular blockade and also the fact that they cannot tolerate inhalation anesthetic agents drugs; all result in their anesthesia being complicated.

In XP patients, using inhalation anesthetic agents such as halothane result in worsening of the symptoms of XP by deranging Nucleotide Excision Repair (NER) in cells; thus general anesthesia using volatile agents should be avoided if possible⁹ and intravenous agents should be used instead. Also due to neuronal and muscular dysfunction in XP patients they are very sensitive to neuromuscular blocking drugs and the use of muscle relaxants should be minimal.¹⁰ In this case having in mind the risk of difficult laryngoscopy and intubation we used LMA and opted for intravenous general anesthesia. Airway manipulation was minute and the surgery was uneventful.

Conflict of Interest

There is no conflict of interest.

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