CASE REPORT

Anesthetic management in a case of xeroderma pigmentosum

Pravin Ubale, MD[1], Namita Baldwa. MD[2], Ketan Katariya[3], Rushikesh Nalavade[3], Momita Mondal[3]

[1]*Associate Professor;* [2]*Assistant Professor;* [3]*Resident*
Dept. Of Anesthesiology, Topiwala National Medical College & BYL Nair Charitable Hospital, Mumbai.

Correspondence: Pravin Ubale, MD, Anand Bhavan, B Building, Apartment No. 16, Topiwala National Medical College (TNMC) & BYL Nair Charitable Hospital Campus, Mumbai Central, Dr. A. L. Nair Road, Mumbai, Maharashtra 400008, (India); Tel: +9322211472; E-mail: drpravinubale@gmail.com

ABSTRACT

Xeroderma pigmentosum (XP) is a rare autosomal recessive disease, which is characterized by hypersensitivity of the skin to ultraviolet (UV) radiation and progressive neurological complications. Inhalational anesthetic agents are avoided due to the possibility of inducing DNA damage and minimum use of muscle relaxants is done intraoperatively. These patients may have a potential difficult airway. We report a 6 yrs old male child who was a known case of xeroderma pigmentosa scheduled for a dental extraction under general anesthesia. The uneventful course of anesthesia in this case was due to a thorough systemic evaluation and careful anesthetic plan.

Key words: Xeroderma pigmentosum; General anesthesia; Total intravenous anesthesia

Citation: Ubale P, Baldwa N, Katariya K, Nalavade R, Mondal M. Anesthetic management in a case of xeroderma pigmentosum. Anaesth Pain & Intensive Care 2015;19(4):524-526

INTRODUCTION

Xeroderma pigmentosum (XP) was first described in 1874 by Hebra and Kaposi. In 1882, Kaposi coined the term for this condition, referring to its characteristic dry, pigmented skin. XP is a rare disorder transmitted in an autosomal recessive manner. It is characterized by photosensitivity, pigmentary changes, premature skin aging, and malignant tumor development.[1] The manifestations are due to a cellular hypersensitivity to UV radiation resulting from a defect in DNA repair. The basic defect in XP is in nucleotide excision repair (NER), leading to a deficient repair of DNA damaged by UV radiation.[2] An XP variant has also been described. The defect in this condition is not in NER, but is instead in postreplication repair. In the XP variant, a mutation occurs in DNA polymerase η.[3-4] We hereby report a case of dental extraction in a 6 yrs male child under general anesthesia using propofol as an induction agent as well as maintenance agent with minimum use of muscle relaxant with no perioperative complications.

CASE REPORT

A 6 years, 15 kg male child was referred to our institute for dental extraction under general anesthesia. Child was a known case of XP but was not on any medication (Figure 1).

He was also a known case of attention deficit hyperactive disorder (ADHD). Parents also gave history of developmental delay in child and history of allergy to sulpha group of drugs. In view of multiple tooth caries, patient was posted for dental extraction under general anesthesia.

On the day of surgery, a high risk consent was obtained from the parents. All difficult airway accessories including pediatric size laryngeal mask airway and Proseal® laryngeal mask airway were kept ready. Lignocaine sensitivity test was done prior to surgery as there is routine use of lignocaine for infiltration during extraction. After taking the patient on the operating table, baseline blood pressure (110/70 mmHg), pulse rate (94/min) and oxygen saturation (98%) were noted. Electrocardiogram (ECG), non-invasive blood pressure (NIBP), precordial stethoscope and pulse-oximeter were used for intraoperative monitoring. Peripheral venous access was achieved with a 22G intravenous catheter. Patient was premedicated with inj. glycopyrrolate 0.04 mg/kg, inj. fentanyl 2 μg/kg,

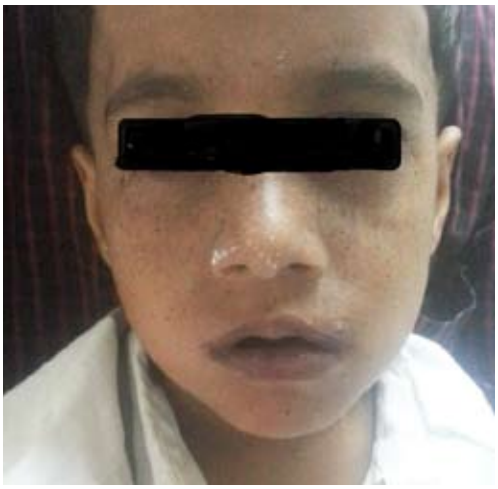

Figure 1: Facial impression showing pigmentation

midazolam 0.03 mg/kg and inj. ondansetron 0.08 mg/kg intravenously. Patient was preoxygenated with 100% oxygen for 3 min and induction was carried with propofol 2 mg/kg followed by succinylcholine 1.5 mg/kg. Nasal intubation was carried out with Portex® uncuffed tracheal tube of size 5 mm internal diameter. Throat packing was done. Anesthesia was maintained with propofol infusion and oxygen 50% + nitrous oxide 50% with Jackson Rees pediatric circuit. The child was allowed to breathe spontaneously with periodic manual assistance. Intraoperative hemodynamic parameters remained stable throughout. Surgery lasted for 1 hour. Ten minutes before the end of the surgery, propofol infusion was terminated, throat pack was removed, suction done and the patient was allowed to recover. Child was extubated after the child attained full recovery. He was shifted to post anesthesia care unit for postoperative monitoring.

DISCUSSION

Xeroderma pigmentosum (XP) is an autosomal recessive genetic disorder of DNA repair in which the ability to repair damage caused by ultraviolet (UV) light is deficient. In extreme cases, all exposure to sunlight must be forbidden, no matter how small; as such, individuals with the disease are often colloquially referred to as 'Children of the Night'. Multiple basal cell carcinomas and other skin malignancies frequently occur at a young age in those with XP; metastatic malignant melanoma and squamous cell carcinoma are the two most common causes of death in XP victims. This disease involves both sexes and all races, with an incidence of 1:250,000 in the United States. XP is roughly six times more common in Japanese people than in other groups.

Patient suffering from XP may present with many preoperative and intraoperative difficulties for anesthesiologist like facial and oropharyngeal changes leading to difficult intubation, prolongation of neuromuscular effect, epiglottis subsidence during extubation and above all is the effect of inhalation agents on nucleotide excision repair.[5,6-8] In patients of XP general anesthesia using volatile agents should be avoided, if possible, because inhalation anesthetics may worsen the symptoms of XP as reported that volatile anesthetics such as halothane deranged NER in cells obtained from an XP patient[9]. Therefore we preferred TIVA (total intravenous anesthesia) to volatile anesthetic usage. Literature studies show that for XP patients TIVA is more appropriate than anesthesia with volatile agents as a method of choice for general anesthesia [10]. Muscle relaxants should be used minimally intraoperatively with neuromuscular monitoring in patients of XP as these patients are sensitive to muscle relaxants due to the neuronal dysfunction.[10]

CONCLUSION

The present case demonstrates that XP patients can be safely managed under general anesthesia using total intravenous anesthesia. Muscle relaxant should be avoided or used minimally with neuromuscular monitoring.

Conflict of interest: None

Author responsibility: All of the authors were involved in the care of the patient as well as preparation of this case report.

REFERENCES

1. English JS, Swerdlow AJ. The risk of malignant melanoma, internal malignancy and mortality in xeroderma pigmentosum patients. Br J Dermatol. 1987 Oct. 117(4):457-61. [PubMed]
2. DiGiovanna JJ, Kraemer KH. Shining a light on xeroderma pigmentosum. J Invest Dermatol. 2012 Mar. 132(3 Pt 2):785-96. [PubMed] [Free full text]
3. Gratchev A, Strein P, Utikal J, Sergij G. Molecular genetics of xeroderma pigmentosum variant. Exp Dermatol. 2003 Oct. 12(5):529-36. [PubMed]
4. Ortega-Recalde O, Vergara JI, Fonseca DJ, Ríos X, Mosquera H, Bermúdez OM, et al. Whole-exome sequencing enables rapid determination of xeroderma pigmentosum molecular etiology. PLoS One. 2013. 8(6):e64692. [PubMed] [Free full text]
5. Soen M, Kagawa T, Uokawa R, Suzuki T. Anesthetic management of a patient with
6. xeroderma pigmentosum. Masui. 2006 Feb; 55(2):215-7. [PubMed]
7. Masuda Y, Imaizumi H, Okanuma M, Narimatsu E, Asai Y, Namiki A. Anesthesia for a patient with xeroderma pigmentosum. Masui. 2002 Feb; 51(2):169-71. [PubMed]
8. Miyazaki R, Nagata T, Kai T, Takahashi S. Anesthesia for a patient with Xeroderma pigmentosum. Masui. 2007 Apr; 56(4):439-41. [PubMed]
9. Oliveira CR, Elias L, Barros AC, Conceição DB. Anesthesia in patient with Xeroderma Pigmentosum: case report. Rev Bras Anestesiol. 2003 Feb; 53(1):46-51. [PubMed]
10. Hasanoglu A Gücüyener K, Tümer L, Gürsel T.- Association of xeroderma pigmentosum with thrombasthenia. Turk J Pediat, 1996; 38:261-264. [PubMed]
11. Masuda Y, Imaizumi H, Okanuma M, Narimatsu E, Asai Y, Namiki A. Anesthesia for a patient with xeroderma pigmentosum. Masui. 2002 Feb; 51(2):169-71. [PubMed]

★★★★★