

CASE REPORT

Xeroderma Pigmentosum: Tailoring Anesthesia for the 'Vampire Disease'

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ABSTRACT

Background : Xeroderma pigmentosum (XP) is a recessive genetic disorder causing defective DNA repair due to ultraviolet (UV) light exposure. Challenges include difficult airway and prolonged effects of muscle relaxants. No definitive safe anesthetics are established. The case study involves an XP patient undergoing mass excision with general anesthesia.

Case Illustration : A 10-year-old girl with xeroderma pigmentosum was admitted for excision of a mass. The mass was diagnosed as basal cell carcinoma. Ventilation was challenging. Preparation for general anesthesia included careful airway management and ventilation tests. The patient was not premedicated to avoid unnecessary sedation and was covered with a blanket to minimize UV exposure. the patient was administered fentanyl and propofol, titrated to induce sleep while maintaining spontaneous breathing. After confirming adequate ventilation, atracurium (0.25 mg/kg) was given and intubation was performed. Anesthesia maintenance was managed with Total Intravenous Anesthesia (TIVA) propofol and 2% lidocaine infiltration around the lesion. Post-surgery, the patient was extubated awake and responsive. Postoperative analgesia included ibuprofen (10 mg/kg), and the patient was discharged after 2 days of recovery.

Conclusion : Patients with xeroderma pigmentosum (XP) require special considerations for anesthesia to avoid worsening their condition. In the discussed case, TIVA was chosen over volatile anesthetics. Additionally, neuromuscular relaxants were used at reduced doses, and multimodal analgesia was used to minimize opioid use. Further research with larger sample sizes is needed to identify the most optimal anesthetic approach for XP patients.

Keywords: Anesthesia Management; Perioperative; TIVA; Vampire disease; Xeroderma Pigmentosum

INTRODUCTION

Xeroderma pigmentosum (XP) is a genetic disease with recessive inheritance, characterized by impaired DNA repair due to ultraviolet light. As a result, genetic mutations occur, leading to skin cancer.¹ In children, the development of skin cancer progresses rapidly. Most skin cancers occur on the face, head, and neck in the form of basal cell carcinoma and squamous cell carcinoma.

The clinical manifestations of XP vary depending on the specific subtype of the disease, but generally include the early onset of skin lesions due to extreme sensitivity to ultraviolet light.¹⁻² These lesions can develop into more severe conditions such as solar melanoma, pigmentation abnormalities, premature skin aging, and an increased risk of developing neoplasms.² About 20% of cases also show neurological complications, and patients may experience ophthalmological symptoms leading to progressive blindness.³ Given the diverse range of symptoms and potential complications associated with XP, comprehensive management strategies are crucial to mitigate its impact on affected individuals.

Several precautions should be taken during the perioperative period for XP

patients, such as avoiding UV exposure in the ward or operating room, avoiding medications that can exacerbate the condition like halothane, and evaluating neurological disorders.¹ Additionally, there may be difficulties in performing endotracheal intubation and prolonged effects of relaxants due to skin atrophy and joint contractures.³

There is no data on the safe type of anesthesia for this condition. Halothane has shown genotoxic effects in XP patients in vitro and may exacerbate the condition in vivo. Sevoflurane can trigger and worsen neurological conditions in XP patients, but there are currently no recommendations to avoid the use of volatile anesthetics in these patients.¹ In the case study to be discussed, the XP patient was consulted for mass excision under general anesthesia.

CASE ILLUSTRATION

A 10-year-old girl diagnosed with xeroderma pigmentosum since the age of 5 presented to the hospital for mass excision and skin grafting on her right cheek. Pathological examination revealed that the lesion was basal cell carcinoma. The lump appeared four months before hospital admission, progressively enlarging and bleeding easily (Figure 1).

Airway examination revealed a

mass measuring 4x5x3 cm on the right mandible that was irregular and prone to bleeding. There was no neck stiffness, and the patient could open her mouth wider than three fingers. The Mallampati score was 3, and keratotic lesions, nasal deformity, keratoconjunctivitis, and visual disturbances were noted. Ventilation was found to be difficult during this examination.



Figure 1. Clinical presentation

The patient weight was 20 kg with a BMI of 16.5, indicating underweight status. Neurological examination showed no abnormalities or developmental delays. The patient was calm and cooperative during the examination. Laboratory and other supportive tests were within normal limits before anesthesia.

The patient was prepared for general anesthesia, with careful

attention to airway management. Ventilation tests were conducted, and the patient could be ventilated with gauze placed around the facial lesion. The patient was brought to the operating room without premedication to avoid unnecessary sedation. Her body was covered with a blanket to reduce UV exposure.

The operating room was prepared with standard monitoring, including oxygen saturation (SpO₂), capnography (EtCO₂), ECG, and non-invasive blood pressure measurement. An intravenous catheter size 22G was previously placed. The patient received fentanyl 2 mcg/kg and titrated propofol until asleep while maintaining spontaneous breathing. Ventilation was tested again using additional gauze, confirming airway control. Atracurium 0.25 mg/kg was administered, followed by indirect intubation with a 5.5 ETT. Anesthesia was maintained with TIVA propofol 50-100 mcg/kg/min, and 2% lidocaine 5 cc was infiltrated around the lesion. Oxygen:air ratio was maintained at 50:50.

The procedure lasted 120 minutes. Hemodynamics remained stable with systolic blood pressure between 94-114 mmHg, diastolic blood pressure 65-75 mmHg, heart rate 65-96 bpm, oxygen

saturation 97-99%, and EtCO₂ 30-35 mmHg. After surgery, the patient was reversed, extubated while awake, and followed instructions well (Figure 2). Postoperative analgesia was managed with ibuprofen 10 mg/kg. The patient was discharged after two days of postoperative care.

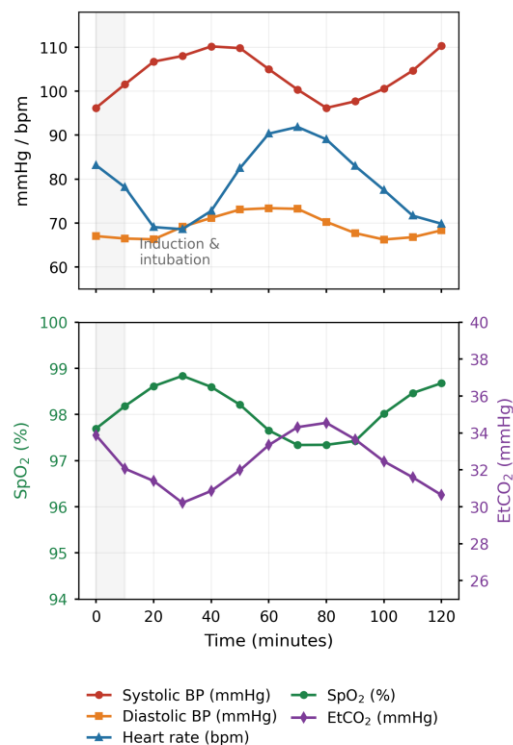


Figure 2. Intraoperative vital sign monitoring across the 120-minute procedure (systolic/diastolic blood pressure, heart rate, SpO₂, EtCO₂).

DISCUSSION

Xeroderma pigmentosum is a rare genetic disorder. This genetic disorder affects sensitivity to UV light. UV light can destroy the DNA of skin cells. Normally, there are genes that can repair

genetic abnormalities, but this does not occur in XP patients. Optimal anesthetic management in these patients needs to be considered.¹

The use of the volatile anesthetic halothane should be avoided. In this case, the anesthesia team was vigilant about potential difficulties with intubation. Ventilation was assessed while the patient was breathing spontaneously.¹⁻² The patient was then given a relaxant and intubated. Some studies have shown that relaxants have prolonged effects.¹ The patient was given a smaller dose of the relaxant, and no prolonged effects were observed in this patient. Several journals suggest that the use of volatile anesthetics should be avoided. Halothane has genotoxic effects. Sevoflurane and isoflurane are associated with the worsening of neurological conditions. Therefore, TIVA (Total Intravenous Anesthesia) becomes the anesthetic choice for XP patients.⁴ In this case, maintenance was done with propofol at 50-100 mcg/kg/min. The most recent update for XP reaffirms TIVA with propofol, ketamine, or dexmedetomidine as the preferred technique, advises avoiding nitrous oxide, and highlights heightened patient sensitivity to benzodiazepines, opioids, and neuromuscular blocking agents, favoring short-acting agents with

neuromuscular monitoring when relaxants cannot be avoided.⁶

In a case report, Miyazaki presented an XP patient with a history of transient neurological symptoms that worsened when volatile agents were used as general anesthesia previously, while the perioperative process proceeded without incident with total intravenous anesthesia (TIVA). On the other hand, Fjouji et al. reported similar findings regarding volatile agents and stated that volatile agents could worsen the neurological status of XP patients, but they did not recommend for or against the use of these agents. Although some animal studies present information on volatile anesthetic-induced neurotoxicity, no human data are available for this effect. Shrestha and Mulimani reported several XP patients who were successfully anesthetized with Propofol as TIVA.⁵

CONCLUSION

The management of xeroderma pigmentosum (XP) patients, especially in perioperative settings, requires meticulous planning and specialized care due to their extreme sensitivity to ultraviolet light and increased susceptibility to various complications, including skin cancers. This case study highlights the importance of careful

airway management, avoidance of UV exposure, and the selection of appropriate anesthetic agents to minimize risks.

In the presented case, a 10-year-old girl with XP underwent successful mass excision and skin grafting for basal cell carcinoma under general anesthesia. The anesthesia plan included the use of fentanyl and propofol, avoiding medications that could exacerbate her condition, and careful monitoring to ensure stability throughout the procedure. The use of gauze around the facial lesion to facilitate ventilation and the avoidance of premedication to prevent unnecessary sedation were critical aspects of the management strategy.

The stable hemodynamics and successful extubation without complications underscore the effectiveness of the tailored anesthetic approach for this XP patient. Postoperative care, including pain management and UV protection, further contributed to the favorable outcome.

This case underscores the necessity of individualized and comprehensive perioperative planning for XP patients, considering their unique vulnerabilities. Future research and data collection on the safest anesthesia practices for XP patients are essential to develop standardized guidelines and improve outcomes for this

rare and challenging condition.

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