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SHORT COMMUNICATION

Dexmedetomidine and ketamine combination for a patient with xeroderma pigmentosa

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Abstract Patients of xeroderma pigmentosa (XP) have increased sensitivity to ultraviolet light and a defective nucleotide excision repair (NER) mechanism in their DNA. Several types of neurological, dermatological, and ophthalmological complications are common in these patients. There is increasing evidence of delayed recovery and worsening of neurological status following general anesthesia in such patients. Some reports have shown uneventful conduct of total intravenous anesthesia in patients of XP. The authors report a case of XP in a young girl for surgery, previously anesthetized with delayed recovery, managed successfully with a combination of intravenous dexmedetomidine and ketamine.

Keywords Dexmedetomidine · Ketamine · Xeroderma pigmentosa

Xeroderma pigmentosa (XP) is an autosomal recessive disorder with increased sensitivity to ultraviolet light and progressive neurological complications due to defective nucleotide excision repair (NER) mechanism in DNA [1]. There are various dermatological and ophthalmological manifestations for which these patients often come for multiple surgeries. Though the anesthetic implications are not clearly known, several case reports have shown worsening neurological conditions including delayed awakening and prolonged neuromuscular blockade [1–3]. We report a patient of XP with delayed recovery during previous exposure to anesthesia who was managed successfully with a combination of intravenous dexmedetomidine and ketamine.

A 16-year-old female patient, a case of basal cell carcinoma scalp with XP, was scheduled for wide local excision with flap cover. She had a previous history of delayed recovery and postoperative mechanical ventilation for 24 h following exposure to general anesthesia 1-year back. We managed her with dexmedetomidine 1 mcg/kg and ketamine 2 mg/kg loading dose followed by 0.6 mcg/kg/h and 0.6 mg/kg/h infusion, respectively (titrated to effect). Bilateral supraorbital nerve block with 0.25 % bupivacaine (1 ml each side) and intravenous paracetamol 15 mg/kg was also given before the start of surgery. In order to avoid exposure to light, except for the surgical site, the rest of the body was kept covered with drapes. The patient was breathing spontaneously with oxygen via facemask and was hemodynamically stable (heart rate, blood pressure, and SpO_2) throughout the surgery that lasted for 100 min. She recovered completely in the immediate postoperative period without any adverse effect later.

The airway may be difficult in these patients because of extensive skin lesions on the face and neck (difficult mask holding) or contractures (limited mouth opening, difficult laryngoscopy). However, in our patient, in spite of widespread skin lesions, we did not anticipate a difficult airway.

It is known that xeroderma pigmentosa is due to a defect in the repair of DNA and is associated with progressive neurological abnormalities. The exact etiology is not

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clearly understood but these are a result of neuronal degeneration, commonly presenting as mental retardation with low intelligence quotient, spasticity, ataxia, sensorineural deafness, and microcephaly in about 20–30 % of patients [4]. In the central nervous system, there is oxidative DNA damage during normal metabolism but most of it is repaired by the NER mechanism. In XP, due to this defective NER mechanism in the DNA, there is a possibility of progressive neuronal damage [5].

In an in vitro study, Reitz and Lanz [6] have shown a possible genotoxic effect of halothane that may contribute to chromosomal defects and disturbances of DNA metabolism in cells. Similar though transitory damage to the DNA has also been shown by sevoflurane and isoflurane in human lymphocytes by Karabiyik et al. [7] in an in vivo study using comet assay. Besides, due to the inherent neurological involvement, the XP patients may possibly be more sensitive to volatile anesthetics as well as benzodiazepines and opioids [8], leading to delayed awakening. It is also hypothesized that XP patients may be sensitive to the muscle relaxants, and therefore some authors have suggested meticulous neuromuscular monitoring in these patients [9].

Total intravenous anesthesia (TIVA) is therefore preferred over inhalational anesthetics and neuromuscular agents for XP patients. In a report by Fjouji et al. [2], a 24-year-old woman with XP underwent general anesthesia for fracture femur surgery after the failure of spinal anesthesia. She had confusion, agitation, and sharpened reflexes after extubation (MRI normal). Later she developed memory disorder and had persisting cognitive decline and spasticity.

On the other hand, Masuda et al. [1] had an uneventful tracheostomy with the use of propofol and fentanyl in a 17-year-old woman with XP. Miyazaki et al. [3] used TIVA without any adverse effect in a case that had transient deterioration of neurological symptoms during previous exposure to volatile anesthetic. Shrestha et al. published a series of three cases of XP managed with TIVA and found increased sensitivity to the synergistic effect of benzodiazepines and opioids in one of the cases. They have recommended the use of non-steroidal anti-inflammatory drugs along with regional nerve blocks as part of multimodal analgesia in patients of XP.

We used a combination of dexmedetomidine and ketamine for TIVA as these two drugs balance the hemodynamic effects of each other (dexmedetomidine lowers the heart rate and arterial pressure induced by ketamine), and maintain spontaneous respiration in clinical doses. Besides, in this case there was no ketamine-induced postoperative delirium, probably because of the use of dexmedetomidine [10]. There was no other post-operative neurological deterioration, suggesting the safety of these drugs in patients with XP.

This case report shows that a combination of dexmedetomidine and ketamine can possibly be used for a patient of XP without resulting in delayed awakening or any neurological sequelae. Nonetheless, it is a guide for further research so that clear insights can be sought about various anesthetic drugs that can be effectively and safely used in these patients by means of further experimental studies.

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