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Dexmedetomidine as an anaesthetic adjunct for total intravenous anaesthesia in patients with xeroderma pigmentosum.

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Xeroderma Pigmentosum (XP) is a rare autosomal recessive disease, caused by a molecular defect in nucleotide excision repair (NER) genes. These patients present with many perioperative anaesthetic challenges like difficult venous cannulation, difficult intubation due to facial and oropharyngeal changes, difficult extubation due to epiglottis subsidence, prolonged action of neuromuscular blocking agents and increased sensitivity to opioids.Harmful effects of inhalational anaesthetic agents on nucleotide excision repair has been proposed. Planned airway management is necessary, short acting opioids with the use of multimodal analgesia is preferred and total intravenous anaesthesia (TIVA) is preferred to inhalational anesthetics. The novel anaesthetic agent Dexmedetomidine, alpha-2 agonist could be a valuable anaesthetic adjunct to TIVA. Besides, it also reduces the induction and maintenance dose of propofol, blunts the airway reflex during intubation and extubation, decreases the requirement of opioids and also helps to enhance the recovery.

We present 3 cases of XP, who underwent surgery using propofol and dexmedetomidine infusion, without the use of muscle relaxants and inhalational agents.

Key words: Dexmedetomidine, Total Intravenous Anaesthesia (TIVA), xeroderma pigmentosum

Introduction

Xeroderma Pigmentosum (XP) is a rare autosomal recessive disease characterized by extreme sensitivity to sunlight and ultraviolet radiation. It is caused by a molecular defect in nucleotide excision repair(NER) genes.[1,2] It presents with prolonged sunburn lesions, pigment changes (freckle-like) on the exposed skin and skin cancer, progressive neurological and neuromuscular complications, and stiffness of the mouth and neck joints.[1,2] Little is known about the optimal anaesthetic management of these patients.

Case Histories

Case 1

A 12 year old male, 30kgs, with xeroderma pigmentosum and basal cell carcinoma of nose, was planned for enucleation of eyeball for fungating mass. (Figure 1) Systemic examination findings and baseline investigations were normal.

After fasting for 8 hours, 2 intravenous cannulas, each of 20G were easily secured in the dorsum of each hand. Patient was shifted to operating room where all the halogenated lights were turned off and base line vitals were monitored.

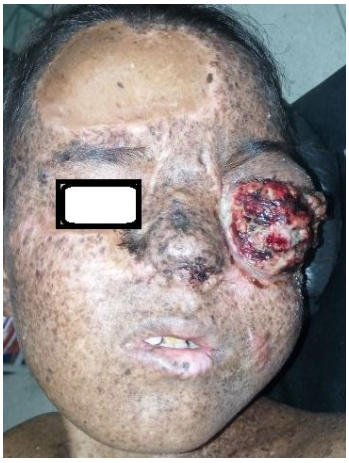

Figure 1.Xeroderma pigmentosa lesions over the face and neck with fungating right eye mass

Dexmedetomidine was given i.v. at 1 mcg/kg and patient was induced with propofol using intermittent boluses of 5mg till the loss of eye lash reflex, with a total of 35 mg.There was adequate

relaxation of jaw and airway was secured with 6mmID cuffed endotracheal tube at first attempt without any significant haemodynamic changes. Intermittent positive pressure ventilation was initiated with tidal volume of 6ml/kg and respiratory rate of 12-16/min titrated to the end tidal carbon dioxide value of 30-35 mmHg. Anaesthesia was maintained with the infusion of dexmedetomidine at 0.5 mcg/kg/hour and propofol at 50-200 mcg/kg/min, titrated to Bispectral Index Score (BIS) of 40-50. Fentanyl 30 mcg, ketorolac 10mg, ketamine 15 mg and paracetamol 500 mg were supplemented as analgesia. The total duration of surgery was 2 hours and throughout the vitals were stable. At the end of surgery, all infusions were stopped and awake extubation was done. His postoperative period was uneventful.

Case 2

A 32 years male, 57 kgs, with xeroderma pigmentosum, was planned for wide excision of squamous cell carcinoma of lower lip, and had generalized widespread macular and hypopigmented skin lesions. (Figure 2) Other systemic examination and baseline investigations were normal.

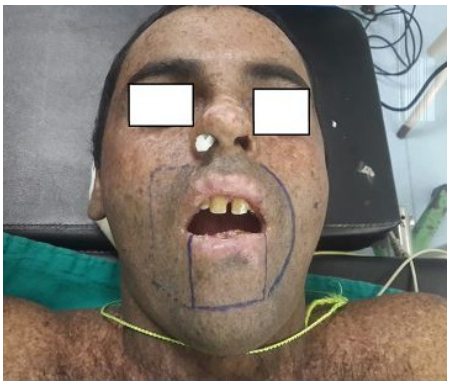

Figure 2. Xeroderma pigmentosa skin lesion with squamous cell carcinoma of lower lip

After adequate fasting, 2 intravenous cannulas, one 18G and the other 20G, were easily secured. Patient was shifted to dark operating room and base line vitals were monitored. As in the first case, dexmedetomidine was given intravenously. Intermittent propofol boluses of 10 mg i.v. was given till the loss of eye lash reflex, up to a total of 150 mg. He was intubated with 7.5 mm ID ETT on first attempt without any significant haemodynamic changes and was ventilated with 6ml/kg and respiratory rate of 10-12/mintitrated to the normal endtidal carbondioxide value. Anaesthesia was maintained with the infusion of dexmedetomidine at 0.5mcg/kg/hour and propofol at 50-120 mcg/kg/min, titrated according to the vital parameters. Fentanyl 100 mcg, and paracetamol 1000mg were supplemented as analgesia. The total duration of surgery was 3 hours and throughout vitals were stable. At the end of surgery, all infusions were stopped and awake extubation was done. His postoperative period was uneventful.

Case 3

Same 12 year old first patient presented with ulcerated basal cell carcinoma of nose and planned for wide local incision. (Figure 3) Anaesthetic management was similar as that of first case, except the dose of propofol and dexmedetomidine was titrated based on the vital parameters as in second case. No complications were met in the perioperative period.

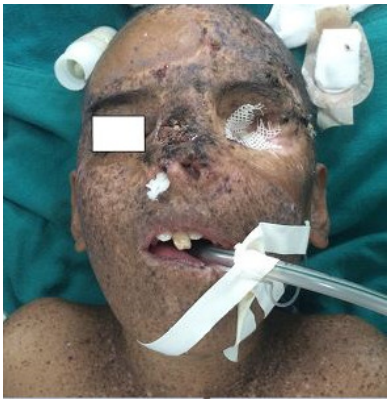

Figure 3. Xeroderma pigmentosa skin lesions over face and neck with right eye enucleation with basal cell carcinoma of nose

Discussion

Xeroderma pigmentosum is a rare autosomal recessive disorder that includes sunlight hypersensitivity, increased risk of skin cancer and neurological manifestations. These patients have a defect in nucleotide excision repair (NER) gene.[1,2] These patients have increased risk of cutaneous neoplasm of the area exposed to the UV rays (face, eyelid, conjunctiva, lips).[2] Patient suffering from

XP present with multiple perioperative challenges for anaesthesiologists.

Intravenous cannulation can be difficult in patients with XP. Facial and oropharyngeal changes can make airway management difficult.[3] No such difficulties were encountered in our cases.Sunlight and halogenated lights can worsen the symptoms in such patients.[3] In patients with XP, general anaesthesia using volatile agents is preferably avoided, as the inhalational agents may worsen the symptoms of XP.[4-6] So in our patients, total intravenous anaesthesia (TIVA) was provided using dexmedetomidine and propofol infusion. Dexmedetomidine reduces the requirement of propofol for induction and both of them can facilitate the intubationwithout the use of a muscle relaxant[7] and such patients are sensitive to muscle relaxants due to the neuronal dysfunction.[6] Dexmedetomidine also blunt the airway reflexes.[8] In all our cases, dexmedetomidine was given at 1 mcg/kg over 10 minutes, followed by bolus doses of propofol and after adequate jaw relaxation, patients were intubated without use of a muscle relaxant and the change of haemodynamic parameters during intubation was not significant. Intraoperative infusion of propofol and dexmedetomidine were titrated targeting BIS of 40-60, however BIS was not available in the second and third cases and the dose adjustment was based on the vital parameters. Short acting opioid fentanyl was used for intraoperative analgesia as such patients are opioids sensitive.[9] Multimodal analgesia is preferred, thus ketorolac, ketamine and paracetamol were used.[9] At the end of surgery, infusions were stopped and patients were extubated. During the extubation, one should be careful as there can be subsidence of the epiglottis leading to airway obstruction.[3] However no such problem was noted in our cases.

Dexmedetomidine is a novel alpha-2 agonist with multitude of beneficial effects as an anaesthetic adjunct. It reduces the induction and maintenance dose of propofol, blunts the airway reflex during intubation and extubation, decreases the requirement of opioids and also helps to enhance the recovery.[8]

We demonstrated the successful use of dexmedetomidine as an anaesthetic adjunct to TIVA, guided by BIS and vital parameters monitoring for anaesthetic management of a patient with xeroderma pigmentosum. To the best of our knowledge, there are few reports of the safe use of dexmedetomidine in a patient with XP. Larger case series to explore the safety and effectiveness of dexmedetomidine as an anaesthetic adjunct for patients with this rare disorder is needed.

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