Case Report

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Anaesthetic Management of Patient with Xeroderma Pigmentosum Posted for Basal Cell Carcinoma Excision

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Abstract

Introduction: Xeroderma Pigmentosum (XP) is a hereditary autosomal recessive disorder due to defect in nucleotide repair genes. In this condition patient might require multiple surgeries for removal of skin and ocular lesion.

Case report: A 17 year old male patient, weighing 43 kg presented with multiple lesions over face since 2 months. He had cutaneous lesions over full body with no major neurological deficit. General anaesthesia was planned. Patient's body protection from artificial light and eye protection given in operating room. Patient Pre medicated with Inj glycopyrrolate, Inj Midazolam and Inj fentanyl. Preoxygenation done with 100% oxygen for 3 min. Induced with Inj Propofol. After elimination of reflexes and reaching enough depth of anaesthesia laryngeal mask airway(LMA) number 3 inserted and good lung ventilation confirmed by chest rise and ETCO2 then LMA fixed in place. Anaesthesia maintained with 50% nitrous oxide in oxygen, IV Propofol with intermittent positive pressure ventilation. Further analgesia supplemented by inj.Bupivacaine 0.25% infiltration locally around wound given. The procedure lasted for 40 min.

Conclusion: Xeroderma pigmentosum patients have risk of worsening neurological disorder with genotoxic drugs and volatile anaesthesia and prolongation of recovery from muscle relaxants. In this case report demonstrated that patient with XP can safely be managed under general anaesthesia with IV propofol, without muscle relaxant and inhalational volatile anaesthetics.

Keywords: Difficult cannulation; Difficult intubation; Inhalation Anaesthetics; Propofol; Xeroderma Pigmentosum.

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Introduction

Xeroderma Pigmentosum is a rare hereditary autosomal recessive disorder due to defect in nucleotide repair genes, Characterized by sensitivity to ultraviolet radiation. UV induced skin tumors and progressive neurological complications. In this condition patient might require multiple surgeries for removal of skin and ocular lesion .Here we are discussing regarding patient diagnosed as Xeroderma Pigmentosum with basal cell carcinoma over forehead and nose posted for wide excision and anaesthetic management.

Case Report

A 17 year old male patient ,weighing 43 kg presented with multiple lesions over face since 2 months, initially smaller in size then progressed to present size 1.5 cm Patient was a known case of xerodema pigmentosa. He had cutaneous lesions such as hyperpigmentation over full body. There was no major neurological deficit and he was posted for excision of lesions.

Examination revealed no pallor, pulse rate of 80/min and blood pressure of 120/60 mmHg, respiratory rate of 18cpm and was afebrile. Systemic examination of cardiovascular, respiratory, central nervous system was normal with per abdomen examination of soft abdomen with no guarding/ rigidity. On airway examination include class 3 of mallampati Laboratory investigations were normal.

Anaesthetic Management

General anaesthesia was planned for the surgery. 20G USG guided IVC secured in right fore arm due to difficult visualization of venous access, preloaded with 500ml RL. Monitoring includes pulse oximetry, ECG, invasive blood pressure, end-tidal carbondioxide. Patient's body was covered to protect it from UV and artificial light in operating room and eye protection given. Patient was Pre medicated with Inj glycopyrrolate 0.2 mg IV, Inj Midazolam 1mg and Inj.fentanyl 80 mcg IV. Preoxygenation done with 100% oxygen for 3 min. Induced with Inj.Propofol 100mg IV.

After elimination of reflexes and reaching enough depth of anaesthesia appropriate laryngeal mask airway(LMA) number 3 was inserted and good lung ventilation was confirmed by chest rise and $ETCO_2$, and LMA fixed in place. Anaesthesia was maintained with 50% nitrous oxide in oxygen, IV Propofol with intermittent positive pressure ventilation. Futher analgesia was supplemented by Inj Bupivacaine 0.25% infiltration locally around wound given. The haemodynamic parameters were monitored which remained stable throughout the procedure. The procedure lasted for 40 min during which 1.5L of crystalloids was given. The patient was extubated when he was awake and obeyed to open eye for command and shifted to recovery room. Postoperative recovery was uneventful.

Discussion

Xeroderma pigmentosum (XP) is an autosomal recessive disease characterized by hypersensitivity to sunlight with neurological abnormalities and high incidence of skin cancer. Xeroderma pigmentosum (XP) was first described in 1874 by Hebra and Kaposi. In 1882, Kaposi coined the term xeroderma pigmentosum. It occurs in about 1:250,000 births in the U.S. and 1:40,000 in Japan could become common in racial groups in which there is consanguinity (1-5).

Patient with from XP many require proper shelding from light, avoidance of drugs which are genotoxic. Difficulties for anaesthesiologist like facial and oropharyngeal changes leads to difficult intubation, prolongation of neuromuscular effect, inhalation agents on nucleotide excision repair.

In one of the case report for elective surgery posted for excision of facial mass in the face, General anaesthesia using LMA, 2% sevofluran as a maintenance without any muscle relaxant.[6]

In one more case report, a case of XP with femoral neck fracture posted for surgical fixation, General anaesthesia performed after failure of spinal anaesthesia, used sevoflurane as a maintenance, all parameters were stable during procedure, after extubation patient had confusion, psychomotor agitation, sharpened reflexes. Here it was proven that sevoflurane had a deleterious effect on the neurological status of this patient.[1]

In another case report , patient with XP underwent laproscopic cholecystectomy had experienced transient worsening of neurological symptoms after anaesthesia with volatile agents. Hence they have used total intravenous anaesthesia(TIVA) for next surgery. Here they have proven that TIVA is better than volatile agent as a method for general anaesthesia for patient with XP.[2,3]

Patients with XP are more sensitive to muscle relaxants due to the neuronal dysfunction hence minimum use of muscle relaxants is recommended under the monitoring of neuromuscular blockade.[7]

Conclusion

Xeroderma pigmentosum patients have risk of worsening neurological disorder with genotoxic drugs and volatile anaesthesia and prolongation of recovery from muscle relaxants. At present there are no recommendatios to avoid the use of volatile anaesthetics. In this case report it was demonstrated that patient with XP can safely be managed under general anaesthesia with IV propofol, without muscle relaxant and inhalational volatile anaesthetics.

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