Anaesthetic Implications in a Case of Indian Patient of
Xeroderma Pigmentosum

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ABSTRACT

Xeroderma Pigmentosum (XP) is a rare autosomal recessive disease which causes skin pigmentation with precancerous lesions, neurological abnormalities. It is a defect in nucleotide excision repair (NER) mechanism. Here we are presenting the anaesthetic implications in a case of eight year old male patient suffering from XP presented for an ophthalmic surgery.

KEY WORDS: Anaesthesia, Xeroderma Pigmentosum, Inhalation Anaesthetics, Propofol
INTRODUCTION

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. Patients with XP show a failure to properly repair UV-induced DNA lesions by the nucleotide excision repair (NER) mechanism. It results in premature development of neoplasias due to an exacerbated hypersensitivity to UV radiation. Therefore, patients with XP must avoid exposure to UV-radiation by use of protective clothing, sunscreen and UV-blocking film\(^1\). This report aimed at describing the anesthetic management of a patient with Xeroderma Pigmentosum submitted to ophthalmologic surgery as there is paucity of anaesthetic information from Indian subcontinent for condition.

CASE REPORT

We are presenting a case of eight year old male with normal mental and neuromotor development with Xeroderma Pigmentosum (Figure 1) and extensive facial involvement, submitted to right eye papillomatous lesion excision. After taking the patient on the operation table, proper positioning and limb support was given. Baseline blood pressure (110/70 mm of Hg), pulse rate (94/min) and oxygen saturation (98%) were noted. Electrocardiogram (ECG), non-invasive blood pressure (NIBP), precordial stethoscope and pulse oximeter were monitored. Peripheral venous access was achieved with a 22G catheter. Patient was given premedication with Glycopyrrolate 0.1 mg, Fentanyl 15mcg & Midazolam 0.25mg intravenously. Patient was preoxygenated with 100% oxygen for 3 minutes and Intravenous Propofol was used for induction in incremental concentrations and 20 mg succinylcholine was used for intubation. Intubation was carried out successfully only with Portex soft seal cuffed tracheal tube of 5mm, a guide wire was used to help tracheal tube introduction as intubation was difficult. Anesthesia was maintained with Propofol and oxygen and nitrous oxide in 50:50 proportions with Bain's Circuit. Patient was extubated in the operating room and was sent to the post-anesthetic care unit in good conditions. Intraoperative hemodynamic parameters were stable throughout. For postoperative nausea vomiting, Ondansetron 2mg was given intravenously. He was shifted to post anaesthesia care unit for further observation. Postoperative recovery was uneventful.
DISCUSSION

Xeroderma pigmentosum (XP) is an autosomal recessive disease that is characterized by hypersensitivity to sunlight with a high incidence of skin cancer and exhibits variable neurological abnormalities. First described by Kaposi in 1870 begins in childhood and progresses determining premalignant and malignant lesions that often cause affected individuals to death in early adulthood. 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. Classical types of XP have a defect in nucleotide excision repair (NER). Patient suffering from XP present with many preoperative and intraoperative difficulties for anaesthesiologist like Facial and oropharyngeal changes leads to difficult intubation, prolongation of neuromuscular effect, epiglottis subsidence during extubation and above all harmful effects of anaesthetic drugs such as inhalation agents on nucleotide excision repair. 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. It is well documented in literature that total intravenous anaesthesia (TIVA) is more appropriate than anesthesia with volatile agents as a method for general anesthesia for xeroderma pigmentosum patients. As patients of xeroderma pigmentosum are sensitive to muscle relaxants due to the neuronal dysfunction and muscle so minimum use of muscle relaxants is recommended that too under the monitoring of neuromuscular blockade.
regional anaesthesia can be appropriate should be preferred over general anaesthesia. As important as the anesthetic-surgical aspect that causes the disease, since this type of patient requires frequent surgeries, is the psychosocial impact on the patient and their families. The progression of the disease has a number of limitations to daily activities of children and their parents require a degree of attention and great care. Thus, it is of fundamental importance to preanesthetic visit attentive, and the correct indication of premedication in order to reduce anxiety and stress experienced by these patients.

CONCLUSION

Patients suffering from xeroderma pigmentosa need proper anaesthetic management with no harmful drugs and protective covering to skin with meticulous care. Proper management of Xeroderma pigmentosa patient is incomplete without proper patient and patient’s relative education towards XP.

REFERENCES