LETTER TO EDITOR

Xeroderma Pigmentosum: What Anaesthetist Should Know?

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Sir,

A six year old male child, weighing 18 kg was referred to Department of Anasethesia, Byramjee Jeejeebhoy Government Medical College and Sassoon General Hospitalfor excision of ulcero-proliferative lesion of upper lip. Lesion was 2.5×2 cm in size arising from angle of mouth for last one month (Fig.1). Patient is a known case of Xeroderma Pigmentosum (XP) since one year of age. He had extensive hyperpigmented macular as well as raised skin lesions in sun exposed areas along with diminution of vision in both eyes secondary to corneal opacity and keratomalacia. Physical examination of child was unremarkable. Preoperative blood investigations were within normal limits. Standard preoperative monitors were attached. Intravenous access was secured gently. Patients head, neck, trunk and extremities were covered with linen before turning the operation theatre lights on. Lesion was covered with sterile gauze prior to face mask ventilation to avoid trauma. General anaesthesia was induced with intravenous midazolam 0.5 mg, fentanyl 40 mcg, propofol 40 mg followed by I-gel no. 2 insertion (Fig. 2). Use of muscle relaxant was avoided. Anaesthesia was maintained with O_2 and propofol infusion along with assisted ventilation via circle absorber. Lubricating eye drops were instilled before patching the eyes. Local infiltration of surgical site was done with 0.25% bupivacaine 3 ml. Intraoperative course was uneventful. iGEL was removed after effortless spontaneous breathing. Postoperative analgesia maintained with appropriate dose of diclofenac and tramadol. Paracetamol was avoided as analgesic. Histo-pathology of lesion confirms basal cell carcinoma of lips.

XP is a rare autosomal recessive disease which causes skin pigmentation with precancerous lesions, ocular lesions and progressive neurological abnormalities [1]. These manifestations are due to a cellular hypersensitivity to ultraviolet radiation leading to a defect in repair of DNA by the process of nucleotide excision repair [2]. Dermatological manifestations include erythema, hyperpigmentation, dry pigmented skin, skin atrophy, telangiectasis and skin malignancy at early years of age. This change hinders the visualization of vein leading to difficulty in intravascular cannulation. Gentle application of noninvasive monitors as well as adequate padding during positioning is necessary to prevent skin injury [2]. Topical application of 5-fluorouracil for premalignant lesion may result in bone marrow suppression which warrants cautious use of nitrous oxide, ocular manifestations including photophobia, keratitis, cataract, lid skin atrophy and malignancy [3], lubricating or antibiotic eye drops should be used to protect the eyes before patching. UV-absorbing eye glasses are also available for eye protection. Neurological manifestations include spasticity, ataxia, seizures, peripheral neuropathy, microcephaly, progressive intellectual impairment and diminished tendon reflexes, occur due to neuronal loss, nerve degeneration and nerve

Fig. 1: Child with Xeroderma Pigmentosum having Ulceroproliferative Lesion
Fig. 2: Airway Management with iGEL

demyelization [3]. These changes influence the selection of anaesthetic agents and anaesthesia technique. Requirement of muscle relaxant is less due to neuronal dysfunction and should be used judiciously with neuromuscular monitoring [4]. Inhalational anaesthetic agents and paracetamol should be avoided as they derange nucleotide excision repair in cells and cause worsening of symptoms [5, 6]. Hence, regional anaesthesia is preferred over general anaesthesia and total intravenous anaesthesia should be opted if general anaesthesia is necessary. Progressive skin lesions of head and neck may lead to microstomia and difficult face mask adaption thus increasing the airway difficulty. Psychological and social impact to these patients due to progressive nature of lesion and frequent exposure to surgery and anaesthesia warrants good communication as well as anxiolysis in preoperative holding area. With the knowledge regarding the genetic disorder and preoperative planning and preference to total Intravenous anaesthesia results in a safe perioperative outcome.

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