A child with xeroderma pigmentosum for excision of basal cell carcinoma

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ABSTRACT

Xeroderma pigmentosum (XP) is characterized by hypersensitivity to sunlight, ocular involvement, and progressive neurological complications. 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. Basal cell carcinoma which is rare in children can occur with XP. Though the XP induced changes are predominately dermatologic, pose several challenges in anaesthetic management. Hence, we are reporting a 9-year-old child with XP scheduled for excision of basal cell carcinoma under general anaesthesia.

Key words: *Anaesthetic implications, basal cell carcinoma, xeroderma pigmentosum*

INTRODUCTION

The description of basal cell carcinoma in children is unusual, but it can occur with xeroderma pigmentosum, a rare hereditary autosomal recessive disorder.[1] Xeroderma pigmentosum is characterized by hypersensitivity to sunlight, ocular involvement, and progressive neurological complications.[2] These manifestations are due to a cellular hypersensitivity to ultraviolet (UV) radiation leading to a defect in repair of DNA by the process of nucleotide excision repair (NER).[2]

Though the XP induced changes are predominately dermatological, pose several challenges in anaesthetic management. Here, we are reporting a child with XP scheduled for excision of basal cell carcinoma under general anaesthesia.

CASE REPORT

A female child of 9 years old, diagnosed to have xeroderma pigmentosum 6 years back, presented with wound over the tip of the nose and watering of the eyes on exposure to sunlight. On examination, the child was conscious, cooperative, poorly built, and nourished, presented with generalized bilateral distributed freckles, and hypo pigmented nodules seen over face, trunk, and extremities. Ulcerated nodule of 2×2 cm^2 with crusting seen over the tip of the nose. Hypo-pigmented macules were present over lips. Conjunctiva was congested, and the cornea was hazy. Immature cataract and entropion were present in left eye [Figure 1]. Investigations were within normal limits. Chest X-ray was normal.

General anaesthesia was planned for surgery. After taking consent IV line was cannulated on hand. Preoxygenation was done by holding the mask at a distance in the face. The

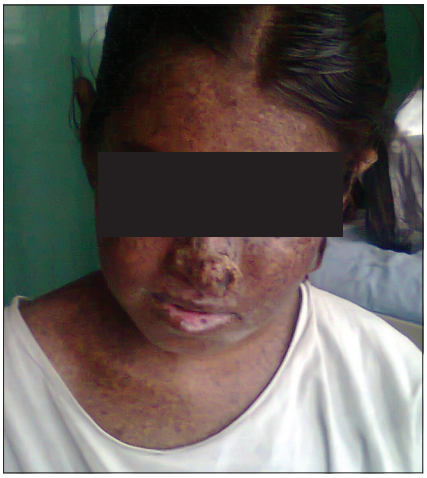

Figure 1: Child with XP and basal cell carcinoma of tip of nose

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child was pre-medicated with atropine 0.02 mg/kg, fentanyl 0.5 ug/kg, and midazolam 0.5 mg/kg intravenously. Once the child was induced with propofol 2 mg/kg, then trachea was intubated with 5.5 no cuffed oral endotracheal tube after suxamethonium 2 mg/kg IV in a single attempt. Anaesthesia was maintained with O_2, N_2O, and inj atracurium 0.3 mg/kg by controlled ventilation using Bains circuit. Local infiltration of surgical site was done with inj bupivacaine 0.5% 3 ml. Intra operative monitoring was done with SPO_2, NIBP, and ECG. Intra operative course was uneventful. Neuromuscular block was reversed with reversal agents and trachea extubated at the end of surgery.

DISCUSSION

Among skin cancers BCC is commonest non melanoma skin cancer and contributes 75% of cancers which occurs in the 7th decade of life in white population.[1] However, sqamous cell carcinoma is commoner than BCC in dark-skinned individuals.[3]

Though it is rare in children BCC may be associated with genetic factors like albinism, bazex syndrome, basal cell carcinoma nevus syndrome, nevus sebaceous, xeroderma pigmentosum (XP), and non genetic factors like severe sun exposure and high dose of radiotherapy.[1] The incidence of XP is 1:250,000 births in the USA, in Japan 1:20, 000.[4] XP is found in all continents and racial groups but rare in blacks.[5] Ultraviolet light found in sunlight damages genetic material (DNA) in skin cells. Normally, the body repairs this damage but in persons with XP, the body does not fix the damage due to molecular defects in genes involved in NER.[2]

The disease is characterized by dermatological manifestations like severe sunburn, persistent erythema, marked freckle-like pigmentation of the face before 2 years age, dry pigmented skin, atrophy, keratosis, telangiectasis, and neoplasm. Patients with multiple primary lesions, with XP will develop skin cancer by 8 years of age.[2] These children may present with ocular changes before skin lesions like photophobia, keratitis, atrophy of the skin of the lids, cataract, and eye tumors. 25-30% of XP individuals develop neurological manifestations in infancy or second decade. The neurological abnormality includes microcephaly, progressive intellectual impairment diminished deep tendon reflexes, sensory neural hearing loss, spasticity, ataxia, seizures, pyramidal syndrome, and peripheral neuropathy. These neurological symptoms are due to loss of neurons, particularly in the cerebrum and cerebellum, primary axonal degeneration in peripheral nerves, and secondary demyelization. The most common causes of death in XP subjects are due to skin cancer, neurological degeneration, or internal cancer.[4]

Definitive treatment of XP is not well established, since DNA injury is cumulative and irreparable. Hence, persons with XP must avoid exposure to sources of UV light and must wear protective clothing, UV-absorbing eye glasses. Topical application of 5-fluorouracil or imiquimod is used for premalignant lesion and surgical excision is done for malignant neoplasm of the skin, tongue, eyelids, conjunctiva, and cornea. Methyl cellulose or quinodine containing eye drops, and bland ointment are used in eye-care.[2]

The major anaesthetic concerns are the psychological and sociological impact on patient and relatives, due to repeated exposure to surgery and anaesthesia, dry, pigmented skin hinders visualization, cannulation and fixation of venous access, and various non-invasive monitoring techniques. XP demands eye care and protection of the patient from artificial light exposure. Well padding of pressure points and movements should be gentle to prevent skin injuries. Head and neck skin lesions, a microstomia and oropharyngeal injury lead to difficult facial mask adaptation, difficult laryngoscopy and tracheal intubation. The presence of variable neurological abnormalities is very challenging in choosing induction agents, muscle relaxants, and regional anaesthesia techniques.[5-7]

There is a scarcity of anaesthesia technique for XP patients in literature search. However, general anaesthesia with TIVA technique, using propofol and fentanyl is reported by Miyazaki *et al.* study.[7] In Masuda *et al.* study[6] avoided nitrous oxide because 5 fluorouracil can cause myelosuppression and avoided inhalational agents such as halogenated anaesthetics cause genotoxic effects and deranged NER in cells. Oliveira *et al.* study showed that minimum usage of muscle relaxants under the monitoring of neuromuscular block is done in patients of XP,[5] due to neuronal dysfunction, skin atrophy, and joint contracture.

Though BCC is local invasive skin cancer can be excised under local infiltration anaesthesia. We successfully managed the child under general anaesthesia using local infiltration around the lesion, minimal dose of atracurium, and avoiding volatile agents in the perioperative period.

CONCLUSION

This case report highlights the importance of understanding the progressive pathophysiology, recognition of complications followed by careful monitoring will influence the outcome of multiorgan, multisystem genetic syndrome, XP in the perioperative period.

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