



CASE REPORT

Management of dental trauma in a child with Xeroderma Pigmentosa



Nidhi Agarwal *, Dipanshu Kumar, Aakansha Sharma, Ashish Anand

Department of Pedodontics and Preventive Dentistry, Institute of Dental Studies and Technologies, Kadrabad, Modinagar, Ghaziabad 201201, India

Received 25 April 2017; revised 31 August 2017; accepted 18 September 2017
Available online 23 September 2017

KEYWORDS

Xeroderma Pigmentosa;
Dental trauma;
Avulsion

Abstract Xeroderma Pigmentosa is a rare dermatological autosomal recessive disorder that manifests itself early in life as severe sunburn usually after a short exposure to sunlight. The prime characteristic features include photosensitivity, hyperpigmentation and ichthyosis in sun exposed areas, and an increase in the risk of basocellular and squamous cell carcinomas and melanomas of the skin and eyes. The case report highlights the preventive treatment options along with all necessary precautions that should be taken to protect the patient from any iatrogenic inadvertent exposures that may be deleterious to his present state. The purpose of the report is also to discuss the important role of dental professionals when dealing with debilitating medical conditions.

© 2017 The Authors. Production and hosting by Elsevier B.V. on behalf of King Saud University. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Background

Xeroderma pigmentosum is a rare genetic disorder that presents itself early in life as a severe sunburn usually after a short exposure to sunlight. It is also characterized by photosensitivity, hyperpigmentation and ichthyosis in sun exposed areas, an increase in the risk of basocellular and squamous cell carcinomas and melanomas of the skin and eyes (Hasan and Khan, 2011). On the other hand, deprivation of sunlight causes skin

ageing, making it parched, coarse, and atrophic (Mareddy et al., 2013).

The oral manifestations of Xeroderma Pigmentosa include erythroplakia, leukoplakia, actinic cheilitis and squamous cell carcinoma of the tip of the tongue and lips (Hasan and Saeed, 2015). Along with cases of scrotal tongue, benign migratory glossitis, chronic desquamative gingivitis (Hasan and Khan, 2011) and keratoacanthoma (Saawarn et al., 2011), it has been also reported that these patients usually have poor oral hygiene habits, a high rate of caries, dental plaque, and periodontal disease (Hasan and Saeed, 2015).

Dental trauma resulting in loss of front teeth brought a 13 year old boy to the department. Severe hyperpigmentation and dry, flaky skin, drew the attention of the dentist to his dermatological condition.

* Corresponding author.

E-mail address: doc.nagarwal@gmail.com (N. Agarwal).

Peer review under responsibility of King Saud University.



Production and hosting by Elsevier



Fig. 1 Pre treatment intraoral appearance of the patient.



Fig. 2 Extra oral skin lesion; 2a) the dry and parched lips and perioral area; 2b) back of the neck; 2c) the arms and hands; 2d) legs showing the skin lesion.

2. Case report

A 13 year old male child reported to the Department of Pedodontics and Preventive Dentistry of the Institute with pain following loss of front teeth due to a fall one week ago. Intra oral examination revealed avulsion of maxillary right central and lateral and left central incisors as shown in (Fig. 1). The overlying mucosa was inflamed and red.

On general examination, the boy appeared to have brownish black pigmentation on the face, neck and exposed parts of the arms and legs as shown in (Fig. 2{a-d}). At some places there were thickened plaques of skin along with wound formation due to extreme dryness.

Pigmentation was scanty on the areas unexposed to sun, like, trunk, groin, thighs and legs. His mother revealed that the child was born normally but started having this pigmentation a few months after birth, which was initially less in extent but progressed with age. He also complained of photophobia and diminished vision. The boy was a resident of a nearby village and went to the local school. He had difficulty in doing homework and understanding texts. His elder brother also



Fig. 3 Post treatment intraoral photograph showing the space maintainer inserted.

suffers with a similar condition but it is less in severity. The children had been shown to a local doctor for this condition at the time it had started appearing. The doctor had advised them to keep the skin completely covered, avoid any sun exposure and apply moisturizing lotion frequently. The parents had never sought any further medical aid for their children due to the poor socio economic condition.

The child was referred to the Department of Pediatrics of the attached hospital from where he was sent to the Department of Dermatology. A provisional diagnosis of Xeroderma Pigmentosa was made. The parents were informed of the child's condition and the treatment plan was explained to them. Consent was obtained from the parents for the same.

A fixed space maintainer was decided to replace the missing avulsed teeth. Ethical clearance for the treatment was obtained from the Institutional Ethical Committee. Denture teeth of adequate mesiodistal width were adjusted and waxed up on the cast in their correct position. Occlusal interferences were removed. The teeth were then joined together from the palatal side by light cured fiber reinforced composite. This space maintainer was then seated in the correct position in the patient's mouth by a self-cure composite resin as shown in (Fig. 3).

3. Discussion

Oral manifestations are rarely seen in Xeroderma Pigmentosa but a patient suffering from this condition might require dental treatment. Dental management of a Xeroderma Pigmentosa patient has some important considerations as there is a possibility of occurrence of cancer in perioral and oral region.

In the present case, a removable partial denture would be the simplest space maintainer that can be given to the patient. It consists of an acrylic denture base with acrylic resin teeth. Allergic reactions to denture bases have been reported due to leaching of monomer in self cure resin appliances (Rashid et al., 2015). Toxic substances leached from the resins can have cytotoxic effects and cause irritability in the mucosa (Mac Cabe and Basker, 1976). Hence, a removable space maintainer was not fabricated for the patient. Rather a fixed space maintainer with fiber reinforced composite and resin teeth was given. Fixed space maintainers are indicated in maxillary anterior region once all the permanent teeth have erupted. Growth in the intercanine width is complete with the eruption of the permanent teeth. Thereafter growth in transverse direction

takes place with growth in the sutures which is more significant posteriorly than anteriorly (Bishara et al, 1997). Acrylic denture teeth are made up of highly cross linked polymethyl methacrylate and these show negligible leaching of monomers in organic solvents (Loyaga-Rendon et al., 2007). Also, the artificial teeth in this case are not in direct contact with the gingival tissues so allergic reactions are not expected. A major advantage of fiber reinforced space maintainer is that no tooth preparation is required so the integrity of the young pulp is maintained. Other advantages include the fact that anaesthesia is not normally required, soft tissues are not disturbed which simplifies impression procedures, and margins are supragingival, facilitating oral hygiene maintenance.

The condition of XP might get aggravated by any kind of exposure to natural or UV light (Park and Dock, 2003). Hence, care has to be taken to avoid contact with all possible sources of light. The light curing unit was used only for laboratory fabrication of the space maintainer and self-cure composite was used for seating the appliance in the patient's oral cavity. Also, exposure of light from other sources was also kept to a minimum.

There is no known cure of Xeroderma Pigmentosa (Park and Dock, 2003). Only palliative management consisting of reducing exposure to sunlight and consistent dermatological care, along with surgical excision of persistent tumors can prolong the life expectancy. Clinical examination should be carried out on a regular basis to monitor the same.

Xeroderma Pigmentosa has not been much reported especially from developing nations like India (Chaudhary et al., 2012). If every case gets reported, it will help in providing a better insight into the incidence and prevalence of the condition in rural and urban areas. Proper counseling which involves genetic counseling seems to be an important aspect (Wayli, 2015) related to this disease, as an increased incidence of consanguineous marriages has been reported to be associated with it.

4. Conclusion

A disease like xeroderma pigmentosum can go undetected by parents of rural areas because of the unawareness and lack

of knowledge related to this condition. Timely management, proper counseling and spread of awareness can help save lives. Hence, the significance of a careful examination and history taking can never be underestimated. Dental practitioners may encounter rare systemic conditions whose management requires a multidisciplinary approach for improving the quality of life of the medically compromised child patient.

Conflict of interest

None of the authors have any conflict of interest.

References

- Bishara, S.E., Jakobsen, J.R., Treder, J., Nowak, A., 1997. Arch width changes from 6 weeks to 45 years of age. *Am. J. Orthod. Dentofacial. Orthop.* (111), 401–409
- Chaudhary, M., Jajoo, S.N., Agarwal, R., 2012. Xeroderma Pigmentosum: a case report of two siblings. *J. Immunodef. Disor.* (1), 1–3
- Hasan, S., Khan, M.A., 2011. Xeroderma Pigmentosum with Desquamative Gingivitis a rare case report and detailed review of literature. *J. Cosmet., Dermatol. Sci. Appl.* (1), 164–170
- Hasan, S., Saeed, S., 2015. Xeroderma Pigmentosum-a rare genodermatosis: overview of literature. *J. Pigm. Disord.* (2), 1–5
- Loyaga Rendon, P.G., Takahashi, H., Hayakawa, I., Iwasaki, N., 2007. Compositional characteristics and hardness of acrylic and composite resin artificial teeth. *J. Prosthet. Dent.* (98), 141–149
- McCabe, J.F., Basker, R.M., 1976. Tissue sensitivity to acrylic resin. A method of measuring the residual monomer content and its clinical application. *Br. Dent. J.* (140), 347–350
- Mareddy, S., Reddy, J., Babu, S., Balan, P., 2013. Xeroderma Pigmentosum: man deprived of his right to light. *Scient. World J.* (2013), 1–8
- Park, S., Dock, M., 2003. Xeroderma Pigmentosum: a case report. *Pediatr. Dent.* 25 (2003), 397–400.
- Rashid, H., Sheikh, Z., Vohra, F., 2015. Allergic effects of the residual monomer used in denture base acrylic resins. *Eur. J. Dent.* (9), 614–619
- Saawarn, N., Saawarn, S., Shashikanth, M.C., Chaitanya, N.C.S.K., 2011. Keratoacanthoma with Xeroderma Pigmentosum. *Int. J. Dent. Clin.* (3), 120–122
- Wayli, H.A., 2015. Xeroderma pigmentosum and its dental implications. *Eur. J. Dent.* (9), 145–148