

Xeroderma Pigmentosum with Melanoma of Face and Its Prosthetic Management

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ABSTRACT

Xeroderma pigmentosum is a rare genetic disorder, characterized by cutaneous, ocular and neurological symptoms. Squamous cell carcinoma and melanoma are also its secondary characters. This case report is about maxillofacial prosthetic management of a 10 years old child presented with xeroderma pigmentosum. The nose of the patient was excised surgically due to melanoma. This case report elaborates the role of prosthodontist and the whole procedure of constructing the nasal prosthesis via conventional technique by using the patient's sibling nasal form as template. Regular follow up revealed marked improvement in esthetics, function and ultimately patient's quality of life.

Key Words: *Xeroderma pigmentosum. Maxillofacial prosthetic. Nasal prosthesis. Melanoma. Squamous cell carcinoma.*

INTRODUCTION

Xeroderma pigmentosum (XP) is an autosomal recessive disorder of DNA repair in which the ability to repair damage caused by ultraviolet light is deficient.¹ It is a genetic defect in which nucleotide excision repair enzymes are mutated, leading to a reduction or elimination of Nucleotide Excision Repair.² If tumour suppressor genes e.g. p53 or proto oncogenes are affected, the result may be skin cancer i.e, squamous cell carcinoma, basal cell carcinoma and melanoma.³ XP is characterized by cutaneous, ocular and neurological symptoms.⁴⁻⁶ The disease typically starts with skin symptoms i.e, photosensitivity, pigmentary changes and premature skin aging. Ocular symptoms i.e, photophobia and conjunctivitis are seen in nearly 80% of XP patients starting early in life.⁷ Later on, tumours may also involve the eyes. Progressive neurological manifestations, including cognitive deterioration, deafness, poor coordination, spastic muscles, and developmental delay, reported in about 20 – 30% of the patients.⁷ The lifespan of a person with this abnormality varies, although most do not live past twenty years of age.⁸ The symptoms that doctors look for range from dryness of the skin, sunburn and blistering to blindness, irregular skin pigment, and cancerous tumours.⁹ Diagnosis is then confirmed by analyzing child's DNA, skin biopsies, eye examinations, and cultures of the skin fibroblasts.¹⁰

The purpose of this case report is to elaborate the role of prosthodontist and fabrication of nasal prosthesis by

a conventional technique to improve the esthetics, function to some extent and quality of life of a 10 years old child with XP in a cost effective manner.

CASE REPORT

A 10 years old child reported to Department of Prosthodontics, Punjab Dental Hospital with xeroderma pigmentosum. Patient presented a history of development of irregular dark spots on the skin of face, hands and chest on exposure to sunlight since the age of 4 years (Figure 1 and 2). These spots gradually increased in size, ulcerated for about 8 months. Patient also complained of progressive photophobia and irritation in eyes. Family history of patient was negative. Diagnosis of XP was confirmed after analyzing DNA samples of the patient in a tertiary care general hospital (Mayo Hospital). Biopsy of the skin also confirmed melanoma of skin. The nose was surgically resected leaving a large nasal defect. Surgical reconstruction was eliminated as the defect was quite large and autogenous reconstruction was extremely complex. Patient was referred from surgical Department of Mayo Hospital for maxillofacial prosthetic rehabilitation of the nasal defect resulting from surgical resection. Child was functionally, esthetically and emotionally debilitated.

There were two prosthetic options for construction of nasal prosthesis i.e, conventional technique and digitalizing imaging technology. Digitalizing imaging technology was eliminated due to its complexity and cost. Conventional wax facial impression and sculpture of nose was given the final form using acrylic resin or silicone material. Acrylic resin was chosen for economy. Retention of the prosthesis was planned using undercuts and spectacles. Implants could not be used for the retention of prosthesis owing to young age of patient and insufficient bone quality and quantity. Treatment plan was discussed with child's parents and informed consent was obtained.

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Figure 1 and 2: Xeroderma pigmentosum on hands and chest.



Figure 3: Wax-up on the cast.



Figure 4: Fabricated nasal prosthesis attached onto the spectacles.



Figure 5: Pre-treatment.



Figure 6: Post-treatment.

Procedure: The defect was generously outlined with an indelible pencil. Patient's skin was boxed to the circumscribed outline with a wax conformer. Impression was taken by loading irreversible hydrocolloid within the wax conformer. The impression was boxed and a

working cast was poured. Nose form was selected by choosing patient's sibling as donor of nose form. Molten base plate wax was poured into the alginate mould of the donor. The solidified wax form was retrieved from the alginate mould and was checked for the contour and size. Wax pattern was carved on the model, the nasal features sculpted in the shape of the nose (Figure 3). The contours and alignment of the sculpted form were checked for esthetics and symmetry on patient's defective area. Shade selection was done by mixing acrylic with different stains to match the patient's skin tone. The wax form was flaked and acrylic was packed by conventional compression molding. Acrylic skin colours were mixed in acrylic resin powder prior to packing of the mold for internal staining. Nasal prosthesis was again painted after curing and tinted with external acrylic paints to exactly match the skin of the patient. Retention of prosthesis was gained by attaching to the pair of glasses with the help of auto polymerized acrylic resin (Figures 4, 5 and 6). Patient was given instructions for maintenance and placement of prosthesis. Patient was scheduled for post-insertion follow-up after every 3 months to ensure improved esthetics, function and health of the tissues. Patient was found to be very satisfied with function and esthetics of prosthesis.

DISCUSSION

Construction of satisfactory nasal prosthesis is dependent on many factors i.e, the material, retention and esthetics.^{8,9} Retention is one of the most important considerations in fabricating a successful nasal prosthesis.⁴ Different methods based on mechanical devices or adhesives are discussed in the literature.⁸ Mechanical devices such as eye glasses are not useful in patients with a flat residual tissue bed and gravity may cause vertical displacement.⁹ Adhesives may be irritating and can damage the thin margins of prosthesis during removal.⁷ In the last two decades, osseo-integrated implants have been used for the support of nasal prosthesis.⁵ However, they cannot be used in children⁶ and unpredictable tumour sites.¹⁰

In the present case report, there are particular challenges owing to the young age, progressive nature of xeroderma pigmentosum and low socioeconomic background of the patient. Therefore, the decision was made to construct the nasal prosthesis by a conventional technique where the wax pattern was fabricated from the impression of Patient's sibling. Implants couldn't be used for retention of prosthesis due to insufficient quality and quantity of bone and also due to progressive nature of disease. The prosthesis was fabricated from acrylic resin, which is cost-effective, tissue tolerant and reduces time spent in carving and finishing in comparison to silicone material.⁹ Pair of

glasses were used as means of retention because it also masks the striking appearance of xeroderma pigmentosum and was more aesthetically acceptable in this particular case. This technique relies upon skill and individual ability of both the clinician and the technician. The nasal prosthesis which was made for this patient had good aesthetics and it went unnoticed in public, thus allowing him to go about life without drawing attention to his oro-nasal defect.

Maxillofacial prosthetic restoration of nasal defect of a child with progressive medical condition is a challenge towards pleasing esthetics and function. Latest advancement in dental materials have brought a revolution but conventional techniques still come to our rescue in unfavourable anatomic conditions. Above mentioned procedure is a simple, tissue tolerant and cost-effective method for rehabilitation of such defects. It can help in improving the esthetics, function and ultimately patient's quality of life instantaneously.

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