CASE REPORT

Prosthodontic management of an albinism patient-dental implications and management

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SUMMARY

Albinism is a congenital hypopigmentary disorder. Albinism is due to the dysfunction of the melanin-producing cells (melanocytes) resulting in defective production of melanin from tyrosine through a complex pathway of metabolic reactions. Little is known about the varied dental features that albinism presents with. This case report summarises the features encountered in albinism, the different oral findings available in the literature and also presents a case of an albinism patient treated with fixed partial denture and the precautions to be exercised for such patients.

BACKGROUND

Albinism consists of a group of genetically inherited autosomal recessive conditions which are typically characterised by a congenital reduction or absence in melanin pigment biosynthesis which gives the natural colour in the skin, iris of the eyes and hair.¹ Albinism affects men and women, and is apparent from birth. Traditionally, albinism has been classified according to clinical phenotype, and the two main categories are oculocutaneous albinism (OCA) types 1, 2 and 3, and ocular albinism. All types of albinism have some lack of pigment, but the amount varies from type to type. A person with albinism may have one of the following symptoms:¹⁻⁴

- ▶ Absence of colour in the hair, skin or iris of the eye;
- ▶ Lighter than normal skin and hair;
- ▶ Patchy, missing skin colour;
- ▶ Crossed eyes (strabismus);
- ▶ Light sensitivity (photophobia);
- ▶ Rapid eye movements (nystagmus);
- ▶ Vision problems, or functional blindness;
- ▶ Severe gingivitis;
- ▶ Oral mucosal ulceration and
- ▶ Periodontal diseases.

A dentist can play a significant role in successfully treating albino patients if he/she has basic knowledge about the symptoms of albinism. This article aims to present characteristic dental features of an albino patient and treatment modifications in prosthodontics to manage the same.

CASE PRESENTATION

A 25-year-old woman reported to the department of prosthodontics with the complaint of missing anterior teeth, lost due to trauma (figure 1). Systemic examination revealed absence of colour in skin, eyes and iris of the eye. Apart from this, the patient presented with photophobia, nystagmus

(rapid eye movements) and decreased visual acuity suggesting oculocutaneous type of albinism. On taking history it was found that she is suffering from congenital albinism. Her family history revealed consanguineous marriage of her parents and the younger sibling also having similar problem. On dental examination she suffered from generalised gingival inflammation (gingivitis).

INVESTIGATIONS

- ▶ Albinism is present at birth, and it is usually diagnosed based on the infant's appearance. The most definitive test in determining the albinism type is family history. Genetic sequence analysis is done to find out the type of albinism occurring in family. In the presented case the family history was consanguineous marriage and another sibling suffering from the same.
- ▶ Some physicians believe that a bleeding time should be obtained in all albino persons. Blood examination is done for determining bleeding time to rule out Hermansky-Pudlak syndrome (HPS) or Chediak-Higashi syndrome⁵ (CHS; associated with albinism). If HPS⁶ is suspected, bleeding time, platelet aggregation and platelet electron microscopy is necessary (they have a greater tendency to have bleeding disorders). If CHS⁷ is suspected, a haematologist should evaluate polymorphonuclear leucocyte function. In this case haematological test returned normal values.
- ▶ Hair bulb assays help to indicate the status of tyrosinase activity (hair bulbs are taken from the scalp). This test was not available in our region.
- ▶ An ophthalmologist should perform an electroretinogram test, which can reveal vision problems related to albinism. A visual evoked potentials test can be very useful when the diagnosis is uncertain.⁸

DIFFERENTIAL DIAGNOSIS

Albinism is a congenital disorder. It is a defect of melanin production that results in little or no colour in the skin, hair and eyes. Discoloration of the skin is also seen in leukoderma which is a cutaneous acquired condition wherein the loss of pigmentation is patchy. A related disease is Vitiligo, in which localised areas of the skin lack pigment and resemble that of an albino, while elsewhere on the body the pigmentation is normal.

Also there are several syndromes of albinism associated with systemic pathology. These include

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Figure 1 Preoperative photograph of the patient.

CHS, HPS, Griscelli syndrome, Waardenburg syndrome type II,³ Elejalde syndrome and Cross-McKusick-Breen syndrome.⁹

TREATMENT

There is no treatment or cure for albinism. Albinism does not alter life expectancy or have other serious health effects. Treatment depends on the severity of the disorder. The goal of treatment is to relieve symptoms. In the present case, the patient had existing generalised gingivitis so thorough oral prophylaxis was performed and oral hygiene maintenance instructions were given along with prescribing hexidine mouthwash. Meanwhile a temporary removable partial denture was given to the patient. After improvement of gingival condition patient was advised for fixed prosthesis.

Tinted eyeglasses were used during tooth preparation to reduce photophobia from dental spot light.⁹ Tooth preparation was done carefully avoiding tissue laceration. Haemostatic retraction cord was placed and impressions were made with elastomeric impression material (figure 2). An All Ceramic restoration was planned because of its tissue friendly nature (figure 3). The restoration was cemented with dual-polymerised resin cement, Variolink II (Ivoclar Vivadent). Adequate oral hygiene instructions were given for maintenance of fixed partial denture to the patient. Patient was educated regarding the importance of visiting an ophthalmologist, dermatologist and dentist periodically.

OUTCOME AND FOLLOW-UP

The patient is asymptomatic and on regular follow-up visits.

DISCUSSION

It is believed that lack of melanin pigment in periodontal tissues might influence periodontal disease progression in albinos but Champsaur¹⁰ found that albinism does not represent a clinical risk factor in the pathogenesis or exacerbation of periodontal diseases for these individuals.

Figure 2 Elastomeric impression of both the arches.

Figure 3 Postoperative photograph of the patient.

Varied dental features have been reported in albinism patients. Enamel hypoplasia has been reported in brothers with OCA in both primary and permanent dentition.^{9 11} In another case of OCA the patient had an upper maxillary lateral incisor showing features of both dens invaginatus (dens in dente) and dens evaginatus, which is a rare phenomenon.¹² Maurer *et al*¹³ found that albinos in their study presented with excessive gingival bleeding, epistaxis and prolonged oozing from cuts and bruises from early childhood.

HPS can result in variable bruising, epistaxis, gingival bleeding.¹⁴ Hattab and Amin¹⁵ reported a case of two brothers with concurrence of Papillon-Lefevre syndrome and type I OCA. An increased vulnerability to severe periodontitis can be seen in CHS which manifests as early-onset periodontitis on a severe localised or generalised basis with premature exfoliation of both dentitions.¹⁶ Also severe gingivitis, ulcerations of mucosa, tongue and hard palate are seen.¹⁶ In CHS OCA is prominent with photophobia, severe gingivitis and oral mucosal ulcerations and periodontal disease.^{3 4}

Wood reported a case of OCA with actinic cheilitis and chronic periodontitis.¹⁷ Anyone who has suffered excessive exposure to the sun can develop actinic cheilitis, but fair-complexioned people, especially people affected with albinism, are particularly at risk.¹⁸ In our case we did not encounter any such prominent findings except for gingivitis.

Learning points

- ▶ Albinism patients should be educated to visit the dentist regularly for oral hygiene maintenance as they are prone for recurrent gingivitis.
- ▶ Adequate oral hygiene instructions should be given to the patients for the maintenance of the restoration.
- ▶ Oral tissue trauma and bleeding should be avoided. Haemostatic retraction cord should be placed to prevent bleeding.
- ▶ Patients should be educated to see a dermatologist regularly to be screened for skin cancer.
- ▶ Patients should be encouraged to consult ophthalmologist as he plays an important role in detecting albinism because most forms of albinism present with ocular features as the primary morbidity.

Competing interests None.

Patient consent Obtained.

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