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DENTAL CONSIDERATIONS OF ALBINO: A CASE REPORT

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Abstract:
Albinism is been characterized by congenital hypopigmentary disorder. Dys-functioning of Melanocytes results in defective production of melanin. It is a complex disease with unknown exact etiopathogenesis. This case report summarizes the features encountered in albinism, the different oral findings available in the literature
Keywords: Albinism, hypo pigmentation, melanin disorder

Introduction:

Albinism is a group of genetically inherited autosomal recessive condition that is specifically characterized by a reduction or absence in melanin pigment biosynthesis since birth.[1]It is classified according to clinical phenotype, and the two important types are oculocutaneous albinism (OCA) having subtypes as type 1, 2 and 3 and ocular albinism.

Type I albinism is due to defect in tyrosinase gene which leads to reduction in melanin synthesis. Vision defect in type I candidates are not correctable with refraction. Type II albinism is due to defect in P gene which encodes for ion channel protein in melanosome. Overall severity in Type II albino individuals is less than Type I. Oculocutaneous individuals have more tendency of sunburn & development of skin cancer. Other extremely rare forms of albinism are rufous & brown 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

- Loss of normal colour of skin, iris, hairs
- Patchy skin with irregular pattern of scattered melanin pigmented macule
- Crossed eyes (strabismus);
- Light sensitivity (photophobia);
- Rapid eye movements (nystagmus);
- Vision problems, or functional blindness;
- Inflammation and ulcerations in oral cavity present frequently
- Poor oral hygiene in most cases leads to periodontal diseases. .

Being a dentist, it is good to have knowledge of various mucocutaneous & other associated features of albinism for proper treatment. Current article aims to present characteristic dental features of an albino patient and treatment modifications to manage the same.

Case report-

A 55-year-old woman reported to the department of oral medicine & radiology with the complaint of missing right mandibular posterior teeth (figure1). 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 (figure2). On taking history it was found that she is suffering from congenital albinism. Her family history revealed consanguineous marriage of her parents and her younger sibling also have similar problem. On dental examination she suffered from generalized gingival inflammation (gingivitis) with generalized attrition. (figure3)

Investigations

- Albinism is congenitally present, and it is diagnosed based on the infant's appearance. The diagnostic 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 type 6 of Hermansky-Pudlak syndrome is suspected, bleeding time, platelet aggregation and platelet electron microscopy is necessary (they have a greater tendency to have bleeding disorders). If type 5 of Chediak-Higashi syndrome5 is suspected, a hematologist should check the blood count, mainly the polymorphonuclear WBC's function. In this case hematological 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.

▸ Ophthalmologist should perform an electro-retinogram test, which can reveal vision problems related to albinism. A visual evoked potentials test can be very useful when the diagnosis is uncertain[8].

Treatment

There is no treatment or cure for albinism. Albinism does not alter life expectancy or have other serious health effect. It is more considered to be esthetic variation of skin than normal. In severe cases treatment is required based on the disease severity. In the present case, patient had generalized gingival inflammation with bleeding gums hence oral prophylaxis was adviced followed by use of chlorhexidine mouthwash. Meanwhile a temporary removable partial denture was fabricated for the patient. After improvement of gingival condition patient was advised for fixed prosthesis. Patient was educated to henceforth maintain oral hygiene & the importance of visiting an ophthalmologist, dermatologist and dentist periodically[9-10].

Figure 1: Missing right posterior mandibular teeth.

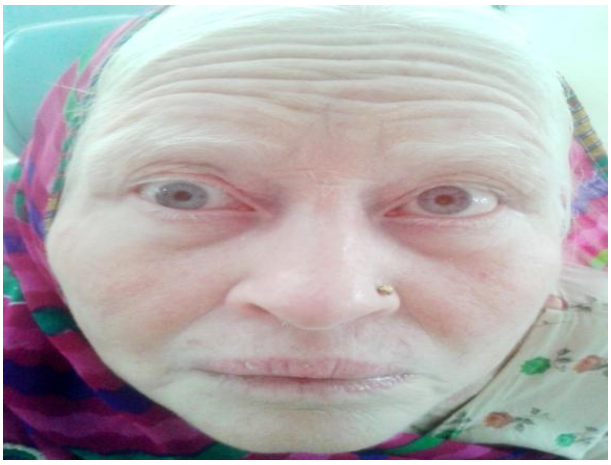

Figure 2: showing involuntary eye movement.

Figure 3: showing generalized attrition and inflammatory changes of marginal and attached gingiva.

Discussion

It is believed that lack of melanin pigment in periodontal tissues might influence periodontal disease progression in albinos but it was found that albinism does not represent a clinical risk factor in the pathogenesis or exacerbation of periodontal diseases for these individuals[10].

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] It was also found that many albino patients 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] An increased vulnerability to severe periodontitis can be seen in CHS which manifests as early-onset periodontitis on a severe localized or generalized basis with premature exfoliation of both dentitions.[16] Also severe gingivitis, ulcerations of mucosa, tongue and hard palate are seen.

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.

Conclusion

Albino 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.

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