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Case Report

Laugier-Hunziker Syndrome - A Rare Cause of Oral Melanosis

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Abstract

Laugier-Hunziker syndrome [LHS] is a rare acquired disorder. It is characterized by diffuse hyperpigmentation of the oral mucosa and longitudinal melanonychia in adults. Appearing as macular lesions less than 5 mm in diameter, LHS is considered to be a benign disease with no systemic manifestation or malignant potential. Therefore, it is important to exclude other mucocutaneous pigmentary disorders that do require medical management. Prompt clinical recognition also forestall the need for excessive and invasive procedures and treatments. In India, the reported cases of this syndrome are very few. In this article, we provide a case report and review of literature on LHS with its differential diagnosis.

Keywords: Benign; Longitudinal Melanonychia; Oral Pigmentation; Acanthosis; Melanosomes

Abbreviations

LHS: Laugier-Hunziker Syndrome; PJS: Peutz-Jeghers Syndrome

Introduction

Pigmentation is frequently encountered in the oral mucosa which may be physiologic or a sign of localized or systemic pathologic condition. A practical approach in a clinical situation is to examine whether the pigmentation presents as focal or as diffuse lesions [1]. Focal lesions usually need an in-depth examination to exclude a melanoma, while diffuse lesions often have no specific histological features and do not generate prognostic perplexity. However, diagnosis of these lesions is important because they could be a sign of diseases with systemic implications such as Peutz-Jeghers syndrome or adrenal insufficiency [2].

Sometimes the clinical behaviour of focal oral pigmentation together with a thorough medical and family history, as well as history of onset, duration, and progression of the pigmentation, guides us to make a suggestive diagnosis. The clinical behaviour of focal oral pigmented lesions ranges from benign, requiring no treatment, to highly malignant. Therefore, biopsy is usually required for accurate diagnosis of a focal pigmented lesion. Histological examination of LHS lesions reveals increased melanin deposition in basal layer keratinocytes as well as an increase in the number of melanophages in the papillary dermis, but there is no increase in the number of melanocytes [4].

One such pigmented lesion which is of interest to the dentist is Laugier-Hunziker syndrome (LHS). It is a rare acquired macular hyperpigmentation of oral mucosa and lips frequently associated with longitudinal streaks of pigmentation of the nails [4]. It was first described in 1970 by Laugier and Hunziker. Since then, the condition has also been detected in other areas with similar histology and the term "idiopathic lenticular mucocutaneous pigmentation" has been used [1].

Case Report

An 18-year old male presented with the history of pigmented spots on lower lip, fingers, and toes. The patient worked in some paint factory since past two years although he had noted the onset of pigmentation when he was 10 years old. Oral mucosa was completely symptom free and the patient was in good health; he did not take any medications regularly and did not smoke. There was no family history of abnormal pigmentation of the skin or oral mucosa. Clinical examination revealed brownish pigmentation on the palm and soles (Figure 1a,1b). Nails of hand and foot showed single longitudinal melanonychia (Figure 1c,1d).

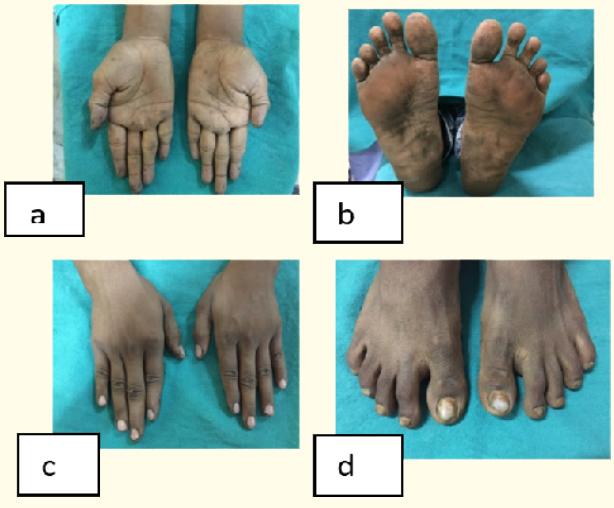

Figure 1a,1b: Palmar and Plantar Surface of Hand and Foot and **Figure 1c,1d:** Nails Showing Longitudinal Melanonychia.

Pinpoint pigmentations were found on the face and several melanin pigmentations can be appreciated in upper and lower lip (Figure 2a,2b). Intra oral examination disclosed diffuse macular brown pigmentation on the right and left buccal mucosa (Figure 2c). There was no pigmentation on the gingiva or tongue. The macules were brown in colour measuring 1-3 mm in dimension.

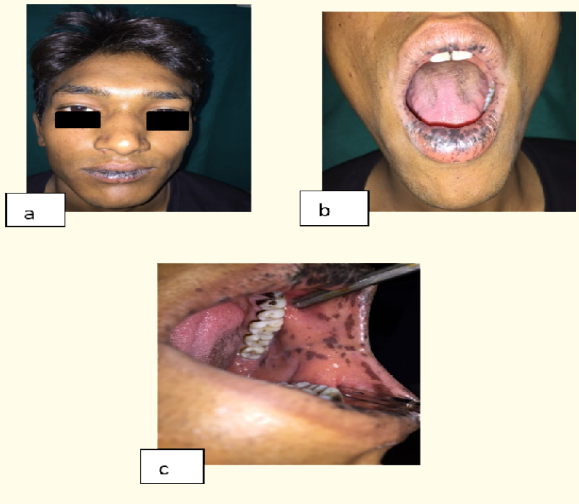

Figure 2a,2b: Extraoral Picture Showing Pinpoint Melanin Pigmentation and Melanin Pigmentation in the Upper and Lower Lip.

Figure 2c: Intraoral Picture Showing Brownish Macule Measuring 1-3 Mm in Diameter.

Laboratory investigations showed a haemoglobin value of 11.2 gm%. ESR was raised to 42 mm/h. Other investigations, including urine cortisol, serum thyrotropin, T3, T4, and adrenotropin were all within the normal range. A biopsy was taken from the oral mucous membrane. The tissue was prepared for routine paraffin processing and haematoxylin and eosin staining. Histology showed stratified squamous epithelium with melanin pigmentations confined to basal layer. The underlying connective tissue showed collagen fibres, fibroblasts and mild perivascular inflammation (Figure 3).

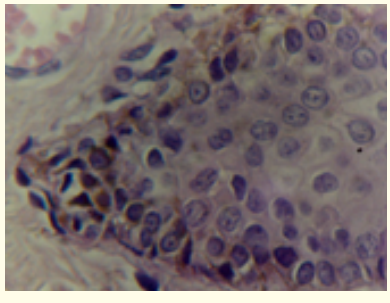

Figure 3: Pictomicroghraph of Buccal Mucosa at 10 X Showing Stratified Squamous Epithelium with Melanin Pigment Confined to Basal Layer.

A final diagnosis of Laugier-Hunziker syndrome was made on the basis of clinical and histological features and absence of systemic involvement. The patient was explained about the benign and sporadic nature of the condition with no systemic involvement. No treatment was performed for the current case and a wait-and-see strategy was adopted.

Discussion

LHS was initially described in 1970 as an acquired, benign skin condition characterized by hyperpigmented macules on the lips and buccal mucosa. It is also associated with longitudinal melanonychia of nails [5].

It is usually manifested in the third to fifth decade of life with a female preponderance in contrast to our case where the patient is young male. The pathogenesis is thought to be linked to a functional alteration of the melanocytes that induces increased synthesis of melanosomes and subsequent transport to the basal cell layers. The etiology of the LHS is still unknown. The pigmentation consists of slate to dark brown lenticular or linear macules, solitary or confluent, with well-defined or indistinct margins. The lesions are located most often on the buccal mucosa and lips and reports

also include pigmentation of the hard and soft palate, the gingival, the palatoglossal arch, the floor of the mouth and the tongue. Nail involvement is present in about half the patients and consists of pigmented longitudinal bands of varying width and intensity in one or more of the fingernails and/or less often toenails as present in our case [6]. The pigmentation may spread from the proximal nail fold into the surrounding skin, which is known as Hutchinson's sign or pseudo-Hutchinson's sign. The hyperpigmentation occurs spontaneously and may progress lowly or remain stable. There are no systemic findings or genetic factors associated with the syndrome.

The mucosal macules of LHS have shown epithelial acanthosis with the pigmentation being localized to the basal layer of the epithelium. The pigmentation is thought to be due to accumulation of melanin in the basal keratinocytes. Various differential diagnoses considered for LHS can be ruled out by the appearance and the etiology for hyperpigmentation (Table1).

Drugs	like tetracyclines, antimalarials, amiodarone, chemotherapeutic agents, oral contraceptives, phenothiazines, azidothymidine, and ketoconazole
Smoking	Seen in anterior gingivaand does not involve the nails
Physiologic (racial) pigmentation of the oral mucosa	Asians, Blacks, and other dark-skinned persons and seen in gingiva
Addison's disease	Immune-mediated destruction of the adrenal gland. Increased level of circulating adrenocorticotropic hormone (ACTH). Oral pigmentation seen as first sign.
Bandler syndrome	Hyperpigmented macules in the hands, nails, and oral mucosa during infancy, as well as intestinal vascular malformation that can cause significant gastrointestinal bleeding.
Mc Cunnie Albright syndrome	Labial and genital pigmentation but is unilateral and does not involve the nails.
LAMB syndrome	Pigmentation of the skin mucosa, atrial and mucocutaneous melanomas, and multiple blue nevi.
Leopard syndrome	Numerous lentigines, electrocardiographic abnormalities, occasional hypertelorism, pulmonic stenosis abnormalities of genitalia, retardation of growth, and deafness [5].
Peutz Jeughers Syndrome (PJS)	Intestinal polyposis and melanotic macules particularly of the face and mouth [8].

Table1: Differential Diagnosis of LHS.

Some case reports have described LHS with oesophageal melanocytosis, actinic lichen planus, hypocellular marrow, and thrombocytopenia, although the relationships with these conditions have not been well established [7].

No treatment is required for this condition since it is not associated with systemic diseases or complications. There have been no reports of malignant transformation therefore all cases must be simply followed-up [9]. Patients who are more aesthetically concerned may choose to have the associated pigmentation removed because of cosmetic disfigurement. A few case studies have demonstrated that cryosurgery, Nd-YAG lasers, and the Q-switched alexandrite laser may be safe and effective options for patients [7]. Recurrence may occur after treatment, but this may be reduced by avoiding exposure to sunlight.

Conclusion

In conclusion, our patient presented a rare syndrome probably not well known among general dentists. Dentists should therefore be familiar with the Laugier-Hunziker syndrome as a benign condition not requiring treatment. When a patient presents with diffuse oral pigmentation, detailed history taking and thorough clinical examination including fingernails will establish the diagnosis and exclude local or systemic diseases requiring medical management.

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