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#### THE LAUGIER-HUNZIKER SYNDROME

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### THE LAUGIER-HUNZIKER SYNDROME

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#### SUMMARY

**Laugier-Hunziker syndrome is a rare, acquired, benign hyperpigmentation of the lips, oral mucosa and nails. Although it is a benign disorder, other pigmentary disorders affecting the oral mucosa and nails must be considered in the different diagnosis. We presented a case of Laugier-Hunziker syndrome, showed clinical and histopathologic features of the disease.**

#### INTRODUCTION

Laugier-Hunziker syndrome is a rare disorder, characterized by macular hyperpigmentation of the lips, oral and genital mucosa with longitudinal hyperpigmented nail streaks(1). Although Laugier-Hunziker syndrome is a benign disorder, other pigmentary disorders affecting the oral mucosa and nails must be considered in the differential diagnosis. We report a female case with the lesions of oral mucosa and nails.

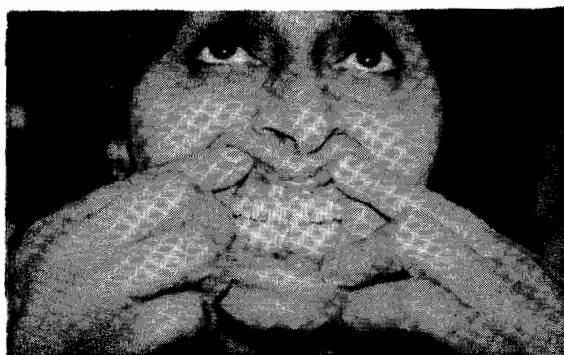
#### CASE REPORT

A 41-year-old female presented with a two year history of nail and periorbital hyperpigmentation. There was no family history of hyperpigmentation and gastrointestinal disorders. The past medical history was unremarkable including chronic medication and smoking.

The physical examination revealed multiple spots on gingiva and buccal mucosa. These pigmented macules were either solitary or confluent. She also had periorbital macular hyperpigmentation (Figure 1). There was no pigmentary changes on genital mucosa, palms, soles and other cutaneous surfaces. Longitudinal hyperpigmented streaks were present on her all fingernails and two toenails (Figure 2).

**Figure 1**

*Macular hyperpigmentation on gingiva and periorbital region*



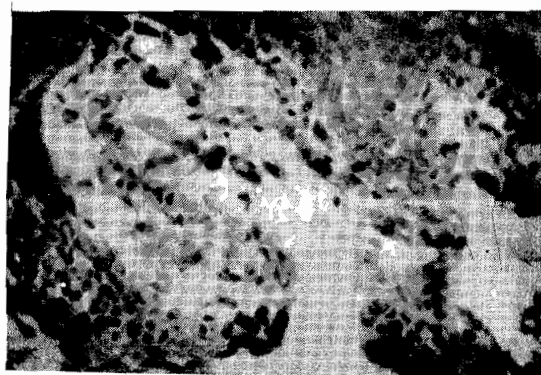
**Figure 2**

*Longitudinal hyperpigmented streaks on nails*



**Figure 3**

*Prominent pigmentation within the basal layer of thickened epidermis (H and E x 400)*



Patient's laboratory tests including ACTH and ANA were in normal ranges. There was also no history of heavy metal exposure. Gastroscopy did not reveal any disease of the gastrointestinal system.

Biopsy was taken from buccal mucosa. The specimen was a tan, brown coloured 3x3 mm punch biopsy. The H and E stained microscopic sections revealed increased melanocytes within the basal cell

layer and intraepidermal spongiotic thickened epidermis. Prominent dilated capillaries were seen in the superficial dermis (Figure 3). Based on clinical and histopathological findings, the patient was diagnosed as Laugier Hunziker syndrome.

## DISCUSSION

Laugier-Hunziker syndrome was first described by Laugier and Hunziker in 1970(2,6). This disorder is an acquired, benign hyperpigmentation of the lips, oral mucosa and nails. Palms, soles and other cutaneous surfaces can also be involved(7). The lesions usually appear during adulthood or elderly. In some cases of Laugier-Hunziker syndrome, other conditions like Sjogren syndrome, Cutaneous Lupus Erythematosus Coombs positive - auto immune haemolytic anemia and inflammatory arthritis have been seen. The significance of comorbid connective tissue disease and Laugier-Hunziker Syndrome is currently unknown. There is no known evidence of a genetic background(3,8). Most of the cases have been reported in Europe(2,8,9). Two articles were published in Turkish literature(10,11). Gurler *et al*(10) reported two cases and one of them had a history of receiving chemotherapy for mammary carcinoma, the other had no history of drug intake. Karakuzu *et al*(11), also reported a case, which was characterized by pigmentations of oral mucosa and nails without coexisting disease or medication.

Although the pathogenesis of Laugier-Hunziker syndrome is not known very well, the possible explanation is that increased synthesis of melanosomes in the melanocytes and subsequent transport to the basal layer cells. However, how the melanocytes are induced to synthesize such a large amount of melanin is not clear(2).

Histopathologic findings are not very specific for Laugier-Hunziker syndrome. Acanthosis, increased number of melanosomes in keratinocytes and increased number of melanophages in the superficial connective tissue are commonly seen features(2,6,8). Our histologic findings were similar to these features.

Lips, oral cavity and hard palate is the most common involved sites. Labial commissures, gums, floor of the mouth, tongue are other locations that are seen less frequently(2,8). Our case had multiple spots on gingiva and buccal mucosa.

In Laugier-Hunziker syndrome, about 60% of cases have nail involvement(4). It is characterized by one or more longitudinal bands of pigment that may vary in width and intensity. These bands occur on the fingernails and/or toenails without nail dystrophy(3). Sometimes Hutchinson's sign may be found. If Hutchinson's sign is present, nail apparatus must be removed and serial sections must be examined to exclude acral lentiginous melanoma(12). Our case had longitudinal hyperpigmented streaks on her nails and Hutchinson's sign was not present.

Other pigmentary disorders of oral mucosa with associated nail involvement must be considered in differential diagnosis. Ethnic pigmentation is seen in dark-skinned persons and most often appear in infancy and puberty(6). Peutz-Jeghers syndrome is an autosomal dominant disorder characterized by pigmented macules of the buccal mucosa, lips, fingers and toes and gastrointestinal polyps. Family history is frequently present in Peutz-Jeghers syndrome. Hyperpigmented macules also occur on the lips and oral mucosa in patients with neuro fibromatosis, Albright syndrome, LAMB, NAME, LEOPARD syndrome and Addison's disease(3). Pigmentary changes can be seen on nails and oral mucosa in lichen planus. Histologic findings of this disease are different from Laugier-Hunziker syndrome. In AIDS, oral pigmented macules and longitudinal melanonychia may be related to systemic drugs, adrenal dysfunction or without precise explanation. Malignant melanoma also must be considered in differential diagnosis(6).

Although, Laugier - Hunziker syndrome does not cause any harm itself, it is important to differentiate from other pigmentary disorders.

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