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OLGU SUNUMU

Laugier-Hunziker sendromu: bir olgu sunumu

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Summary
Laugier-Hunziker syndrome is an idiopathic, macular hyperpigmentation of the oral mucous membranes and the lips, with frequent pigmentation of the nails in a longitudinal pattern. Laugier-Hunziker syndrome is a rare disease with approximately 100 cases reported in the literature. The clinical features consist of dark brown to bluish-black macules mainly located on the buccal mucosa and lips. Involvement of the tongue is exceedingly rare. Herein we report a 25-year-old woman diagnosed as Laugier-Hunziker syndrome, with confluent, hyperpigmented lesions of the tongue, that responded to cryosurgery.
Key words: Cryosurgery, hyperpigmentation, Laugier-Hunziker syndrome

Özet
Laugier-Hunziker sendromu: bir olgu sunumu
Laugier-Hunziker sendromu, idiyopatik, sıklıkla tırnaklarda longitudinal pigmentasyonun gözlendiği, oral müköz membran ve dudaklarda hiperpigmente maküler lezyonlarla klinik seyir gösteren bir hastalıktır. Laugier-Hunziker sendromu, literatürde yaklaşık 100 olgunun yer aldığı ender bir hastalıktır. Klinik bulgular çoğunlukla yanak mukozası ve dudaklarda, koyu kahverengiden mavimsi-siyaha kadar değişen renklerde maküler lezyonlardan oluşmaktadır. Dil tutulumuna ender olarak rastlanmaktadır. Bu makalede, dil mukozasında birleşik, hiperpigmente maküllerin saptandığı, Laugier-Hunziker sendromu tanısı alan ve kriyoterapiye yanıt aldığımız 25 yaşındaki kadın olgu sunulmuştur.
Anahtar kelimeler: Kriyoterapi, hiperpigmentasyon, Laugier-Hunziker sendromu

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Introduction

Also known as idiopathic lenticular mucocutaneous pigmentation, Laugier-Hunziker syndrome (LHS) is an acquired, idiopathic macular hyperpigmentation of the oral mucous membranes and lips, which is frequently associated with melanonychia (1,2). Subjects involved are otherwise healthy. LHS is a rare disease with approximately 100 cases reported in the literature (3).

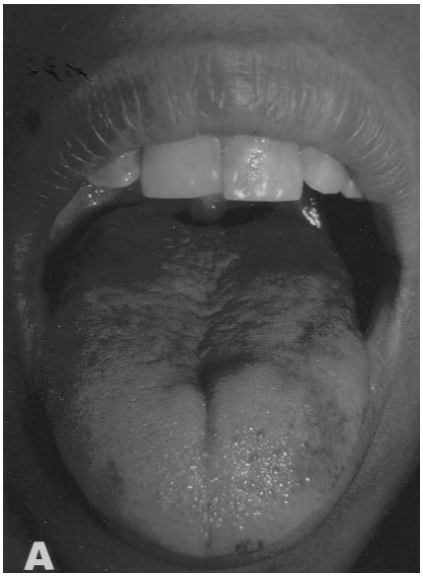
LHS is characterized by the presence of a variable number of asymptomatic, isolated or confluent, slate to dark brown pigmented macules involving the oral mucosa and/or lips, and in 60%, the fingernails (4,5). Herein, we describe a female patient diagnosed as LHS, with confluent, bluish black spots of the tongue, that responded to cryosurgery.

Case Report

A 25-year-old non-smoker Caucasian woman presented with a one year history of progressive hyperpigmented eruption without pruritus or pain on her tongue. She was otherwise healthy and there was no history of medication. The patient denied gastrointestinal or other systemic symptoms. On physical examination we observed well-demarcated, confluent, bluish-black macules irregularly distributed over the lateral margins and tip of the tongue (Figure 1a). There was no other cutaneous or mucosal abnormality.

Laboratory investigations including complete blood count, hepatic and renal functions, serum electrolytes, and urinalysis were within normal limits, and occult stool blood test was found to be negative. A biopsy specimen of a hyperpigmented lesion on the tongue revealed increased basal keratinocyte melanin without expansion of the melanocytic population, superficial pigmentary incontinence with melanophages in the lamina propria and moderate acanthosis (Figure 2). On the basis of clinical and histopathological findings, a diagnosis of LHS was estab-

lished and liquid nitrogen cryosurgery using an open spray, single freeze thaw cycle was carried out. After 2 weeks of treatment a marked improvement of the pigmented lesions was achieved (Figure 1b).



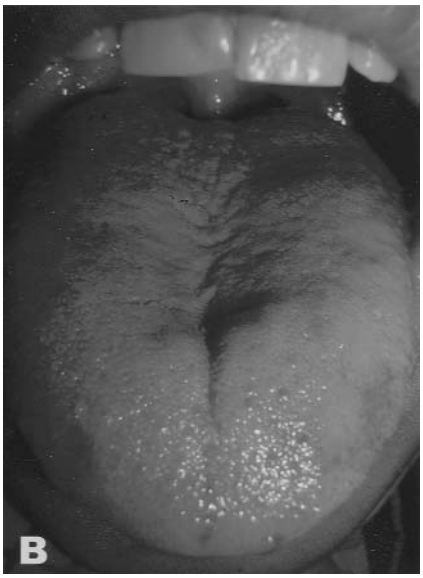


Figure 1a. Confluent, hyperpigmented macules on the lateral margins and tip of the tongue. **1b.** Marked regression of the lesions after cryosurgery

Discussion

Laugier and Hunziker first described five cases of essential melanin pigmentation of the mouth and lips, with longitudinal nail pigmentation (5). The eponymous term Laugier-Hunziker syndrome was later coined to designate the entity.

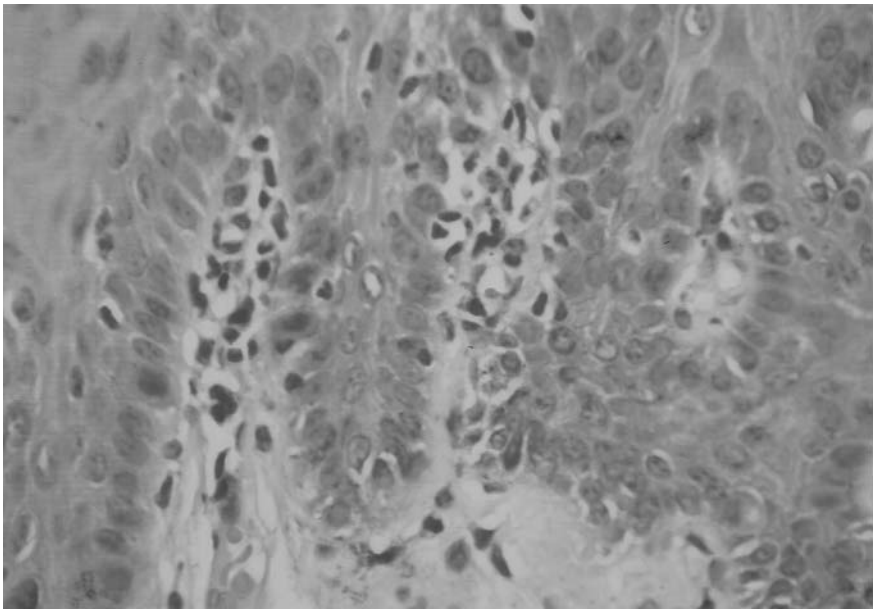


Figure 2. Increased melanin pigmentation in the basal layer of the epithelium, pigmentary incontinence and melanophages in the superficial lamina propria (hematoxylin & eosin stain, X 40)

The onset of the disease is usually between the third to fifth decade of life. There is no sex preponderance (6). Neither familial association nor any associated systemic diseases have been reported, to date. LHS is mainly a disease of Caucasians and most of the reported cases have been from the European countries (7).

The clinical features consist of diffuse or isolated pigmented macules, which have different colors (from gray to bluish-black) with well-defined or indistinct margins. The lesions are most commonly observed intraorally on the lips, the buccal mucosa, the hard and the soft palate, but may also be seen on the gingiva, tongue and the basis of the mouth. Pigmented macules may also occur on the neck, thorax, abdomen, perineal and perianal region, sclerae and esophagus (8,9). Involvement of the tongue in LHS is extremely rare and is usually observed in association with other locations of the oral cavity (1,10). As far as we are aware, this is the second case report of LHS manifesting with only tongue involvement. The incidence of nail pigmentation in LHS is 60%. Baran et al. proposed three types of nail pigmentation in LHS: (i) isolated longitudinal streaks of varying degrees of pigmentation 1 to 2 mm in width, (ii) 2 to 3 mm double longitudinal streaks, and (iii) homogenous pigmentation of the radial or ulnar half of the nail (11). Rarely, the the pigmentation may spread from he proximal nail fold into the surrounding skin (pseudo Hutchinson's sign) (11).

Histological examination reveals increased melanin deposition in the basal layer keratinocytes without an increase in the number of melanocytes, dermal pigmentary incontinence and melanonphages in the upper lamina propia or dermis. Mild to moderate acanthosis may also be observed (5,7). The findings of confluent bluish-black macular lesions on the tongue as well as increased melanin pigmentation in the basal layer of the epithelium, and the presence of melanin incontinence and melanophages in the superficial lamina propria was consistent with a diagnosis of LHS in our case.

The cause of this rare disease is yet unknown. Ultrastructural studies reveal an increase in the number and size of mature melanosomes located in the cytoplasm of basal keratinocytes and dermal melanophages (3). Some authors suggest that functional alteration of the melanocytes in the form of an increase in the synthe-

sis of melanosomes and their subsequent transport to the basal layer cells may give rise to hyperpigmented lesions (12).

The differential diagnosis of LHS includes Peutz-Jeghers syndrome (PJS), Addison's disease, smoker's melanosis, and melanoacanthosis. PJS is an autosomal dominant disorder characterized by hamartomatous gastrointestinal polyposis and melanotic macules, particularly of the face and mouth. Most of the patients have a positive family history of PJS. The polyposis results in episodes of colicky abdominal pain in adolescence or young adulthood. Because of a significantly increased risk of malignancy, comprehensive screening protocols consisting of periodical upper and lower gastrointestinal endoscopies are mandatory (13). Lesions in PJS appear around birth or early childhood, whereas LHS lesions are progressively acquired in young or middle-age adults. Characteristically, in addition to intraoral pigmentation, the lesions are found around the mouth, nose and eyes and on the dorsal and ventral surfaces of the hands and feet in PJS, while the lesions of LHS are mainly confined to the oral mucous membranes and the nails. Appearance of the lesions in later life without involvement of the face and hands, and the absence of a family history and associated symptoms helped us to exclude the diagnosis of PJS in our patient.

In Addison's disease, there is darkening in the areas of trauma, recent scars, points of pressure, areolae, axilla, and perineum. Longitudinal pigmented bands on the nails and diffuse pigmentation on oral mucosal surfaces are observed. Other clinical signs of the disease include fatigue, weakness, loss of weight, hypotension, and gastrointestinal disturbances (14). In our case, lack of related symptoms and normal serum electrolyte levels ruled out this disease. Smoker's melanosis is seen predominantly on the anterior attached gingiva, which is a rare location in LHS. Oral melanoacanthosis shows dendritic melanocytes dispersed throughout the epithelium histologically, which was not detected in our patient (9).

LHS is a benign disease that follows a chronic course with a progressive increase in lesions over years or remain stable. However, spontaneous remission of a case has also been reported in the literature (12). There are paucity of reports of treatment modalities in the literature, probably because of general lack of symptoms in LHS. The reported treatment options include Q-switched Alexandrite laser, Q-switched Nd-Yag laser, and cryosurgery (15,16). Sheridan et al. reported a case of LHS with involvement of the lips successfully treated with cryosurgery (16). Because of the complaint of cosmetic disfigurement, we treated our patient with an eight second, single freeze thaw cycle open spray technique of cryosurgery. A marked improvement was obtained after complete healing in 2 weeks period. To our knowledge, successful treatment of tongue involvement of LHS with cryosurgery has not been reported in the literature.

In summary, we consider that recognition of LHS and differentiation with PJS is important to prevent unnecessary periodical investigations including gastrointestinal endoscopies. We emphasize that cryosurgery should be the first line choice in the treatment of LHS causing cosmetic disfigurement, as a safe, effective and inexpensive method.

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