

A Case of Laugier-Hunziker Syndrome[¶]

LAUGIER-HUNZIKER SENDROMU OLGUSU

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Summary

Laugier-Hunziker syndrome is a benign, acquired and rare syndrome with an unknown aetiology. It is characterized with macular hyperpigmentation of several mucosal and cutaneous surfaces and multiple hyperpigmented bands on the nails.

A case of a 32 year-old female with pigmentations of oral mucosa and nails is reported here. We reported this syndrome, because it is seen very rare.

Key Words: Laugier-Hunziker syndrome

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Özet

Laugier-Hanziker sendromu benign, edinsel ve nadir görülen bir tablodur. Mukozal ve kutanöz yüzeylerde maküler hiperpigmentasyonun yanısıra, tırnaklarda multipl hiperpigmente bandlar görülür.

Burada oral mukoza ve tırnaklarda hiperpigmente lezyonu olan 32 yaşındaki bir kadın hasta sunulmaktadır. Bu sendrom oldukça nadir görülmesinden dolayı bildirimini uygun bulduk.

Anahtar Kelimeler: Laugier-Hunziker sendromu

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Laugier-Hunziker syndrome is a rare, acquired, benign macular hyperpigmentation of the lips, buccal mucosa and particularly palms, soles and other cutaneous surfaces. The nails are often involved with multiple longitudinal hyperpigmented bands (1,2). The sites involved are otherwise healthy (3). The aetiology and pathogenesis are unknown, but Laugier suggests that an enzymatic hyperreactivity in tyrosine-melanin biosynthesis is possibly responsible (4,5).

The syndrome was first described by Laugier and Hunziker in 1970. In this syndrome, the lesions are frequently on lips and buccal mucosa and they are not related to any systemic diseases (4).

Here, we report a female case with the lesions of oral mucosa and nails.

Case Report

Our case was a 32 year-old female otherwise healthy patient, who had mucosal lesions on the left sides of her cheek, tongue and lips and black longitudinal bands on her finger nails. Her symp-

toms had started eight months before her first visit (Figure 1,2,3). Palmoplantar areas, vagina and anal mucosa were not involved. The pigmentation first appeared on her nails. All the laboratory findings were within normal ranges. She had no history of drug intake.

The physical examination was in normal limits. Arterial blood pressure and other vital findings were normal. No gastro-intestinal polyps or other lesions were detected by gastroscopy. There was no family history of pigmentary abnormalities. Two punch biopsies were taken from left buccal mucosa. We found basal hypermelanosis without melanocytic proliferation or cytologic atypia, melanophages in upper dermis and acanthosis in histologic examination (Figure 4). On the basis of clinicopathological findings, a diagnosis of Laugier – Hunziker syndrome was made.

Discussion

Bonnet has first reported physiologic pigmentation of the inner cheeks and lips in a Caucasian



Figure 1. Hyperpigmented lesion on tongue.



Figure 2. Hyperpigmented lesion on buccal mucosa.

man (4); Laugier and Hunziker have described in five cases who had essential melanic pigmentation on the oral mucosa and lips and included their clinical, histological and ultrastructural findings (4,6). Laugier–Hunziker syndrome is very rare. We have found out only one article in Turkish literature. Two cases were reported in that article. One of those had a history of drug but our case had no history of drug intake (7). No sex preponderance and familial factor have been defined(8,9). The syndrome is acquired in early or mid–adult life (3,10). Our case is a 32-year old female without any family history or coexisting disease. These data match the pattern in the literature. The presence of regular acanthosis, basal pigmentation without increase in the number of melanocytes and the presence of melanophages in subepidermal connective tissue are the most constant histopathological characteristics (11). Our histologic findings were similar to these features.

The clinical appearance may be variable and the occurrence of all the pigmented localizations in the same patient is very rare. The most frequent findings are the involvement of the buccal mucosa, pigmentation of the lips, longitudinal melanonychia, pigmented macules around the nails and dark palmo–planter spots, respectively(4). Our case demonstrated mucosal lesions on the left side of her cheek, tongue and lips and black longitudinal bands on her finger nails

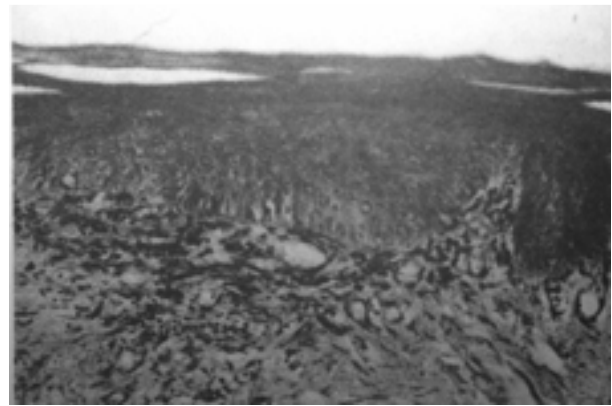


Figure 3. Longitudinal hyperpigmented bands on finger nails.

The importance of this condition is due to its inclusion in the differential diagnoses of pigmentary disorders of the oral mucosa with associated nail involvement. It is important to recognize this acquired benign disorder to avoid unnecessary investigation and treatments (12). The differential diagnosis for the association of mucocutaneous and nails pigmentation includes ethnic pigmentation, Peutz–Jegher's syndrome, lichen planus, human immunodeficiency virus disease, and metastatic melanoma (4).

A pigmentation of proximal nailfold is sometimes seen in dark - skinned persons (13). Our patient was not dark–skinned. In Peutz–Jegher's



Figure 4. Basal hypermelanosis and the melanophages in upper dermis (Hematoxylin-eosin stain; X40).

syndrome, there is hyperpigmentation of fingers and toes and also macular pigmentation of buccal mucosa and lips, in addition to gastro-intestinal system involvement (13). No gastro-intestinal system involvement was demonstrated in our case. Patients with AIDS may have polydactylous involvement and zidovudine also may cause similar pigmentation (13). Our case was HIV negative and the other clinical features of AIDS were not similar to this syndrome. Malign melanoma has a specific histologic appearance unlike this case. No risk of development of oral or subungual malignant melanoma in Laugier-Hunziker syndrome has been reported (4,8).

Therapeutical options for Laugier-Hunziker macules are not discussed in the literature since these are always benign and do not require treatment (11). However, Ferreira et al reported a Laugier-Hunziker syndrome case treated with Q-switched Nd-Yag laser successfully (14).

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