

## Laugier-Hunziker syndrome - Case report\*

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**Abstract:** Laugier-Hunziker syndrome is a rare, acquired disorder characterized by lenticular hyperpigmentation of the oral mucosa and longitudinal melanonychia. We present the case of a 63-year-old female with progressive, asymptomatic hyperpigmentation of buccal mucosa and a 7-year history of hyperpigmentation in several fingernails. Laugier-Hunziker syndrome was diagnosed based on the clinical features presented, dermoscopic findings and exclusion of underlying systemic diseases. Laugier-Hunziker syndrome is regarded as a diagnosis of exclusion. By identifying Laugier-Hunziker syndrome, other, more severe syndromes associated with hyperpigmentations can be excluded, namely Addison's disease and Peutz-Jeghers syndrome.

**Keywords:** Hyperpigmentation; Mouth mucosa; Nails

### INTRODUCTION

Laugier-Hunziker syndrome (LHS) is a rare, acquired disorder characterized by lenticular hyperpigmentation of the oral mucosa and longitudinal melanonychia.<sup>1</sup>

Laugier and Hunziker first described this intriguing phenomenon in 1970 as a condition characterized by adult-onset oral pigmentation and sometimes similar genital lesions.<sup>2</sup> A decade later, Baran noted that this syndrome often includes longitudinal melanonychia.<sup>3</sup> Its etiology is still unknown, and it seems that neither familial predisposition nor systemic and malignant disease is associated with the syndrome.<sup>4</sup>

### CASE REPORT

A 63-year-old, Caucasian female, non-smoker, was admitted to our department for her regular bullous pemphigoid (BP) check-up. The disease treated was in complete remission but she reported progressive asymptomatic blackish pigmentation of both buccal mucosa and mentioned a 7-year history of brownish-black pigmentation of several fingernails. There was no relevant drug history and no other family members had been affected.

Physical examination revealed a regular, brownish, reticular pattern on both sides of her buccal

mucosa (Figure 1). The longitudinal, pigmented band was present on her fingernails and toenails (Figure 2). Potassium hydroxide examination and fungus culture were negative. Digital dermoscopy revealed a pseudo-Hutchinson's sign, with homogeneous brownish and grayish longitudinal bands and lines, with ill-defined margins on the nail plate (Figure 3).

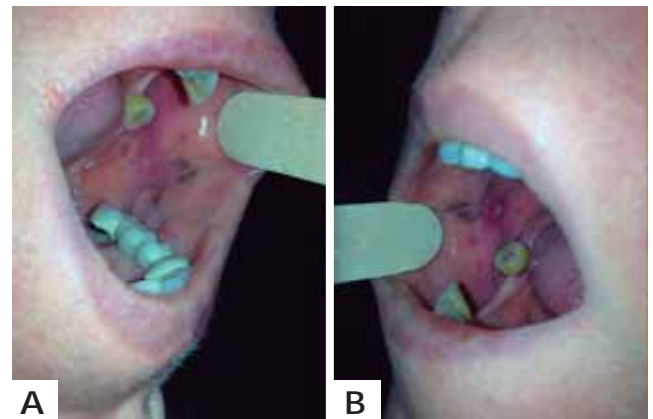


FIGURE 1: A e B. Regular, brownish reticular pattern on the buccal mucosa

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**FIGURE 2:**  
**A.** Homogenous and band-like hyperpigmentation on the patient's fingernails,  
**B.** Longitudinal hyperpigmentation on both first toenails and homogenous pigmentation on all other toenails



**FIGURE 3:** Pseudo-Hutchinson's sign, with homogeneous, brownish and grayish longitudinal bands and lines with ill-defined margins on the nail plate

Laboratory tests were all within normal values. They included the following: erythrocyte sedimentation rate, complete blood count with differential, urinalysis, lipid status, renal and liver biochemistry, serum levels of amylase, lipase, lactate dehydrogenase; tumor markers -carinoembryotic antigens (CEA), alpha fetoprotein antigens (AFP), carbohydrate antigen (CA 19-9); immunology parameters - antinuclear antibodies, cold agglutinins and cryoglobulins.

Chest X-ray and abdominal ultrasound showed no pathological findings.

Colonoscopy was performed to exclude any gastrointestinal disease associated with oral hyperpigmentation. Findings were negative for any tumorous formations in the distal and proximal regions of the colon.

There was no suggestion of adrenal insufficiency, based on the endocrinologist's exam, levels of cortisol and antipituitary antibodies, as well as the adrenocorticotropic hormone (ACTH) stimulation test (Synacthen test).

Our patient declined biopsy of the oral pigmented lesions.

### DISCUSSION

At present, no more than 180 cases of LHS have been reported in the literature, and this mucocutaneous disorder is not believed to be associated with systemic abnormalities.<sup>4</sup>

Although Laugier and Hunziker noted that hyperpigmentations are frequently present on the oral mucosa, pigmented lesions have also been described in atypical locations, such as the neck, thorax, abdomen, pretibial area, sclera, eyebrow site and esophagus.<sup>5</sup> The pigmentations typically develop during early to middle adulthood with a female preponderance, and an overall female to male ratio of 2:1.<sup>6</sup> A mean age of 52 years and median age of 42 years have been reported.<sup>5</sup> The syndrome also involves nail pigmentations, usually vertical, longitudinal bands. Occasionally, pseudo-Hutchinson's signs may be present when hyperpigmentation of the nail bed and matrix reflects through the transparent nail folds, simulating true Hutchinson's signs.<sup>7,8</sup>

Even though our patient declined a biopsy of the oral lesion, the histology by itself was rather unspecific, showing epithelial acanthosis, with pigmentation localized in the basal layer of the epithelium.<sup>6,7</sup>

Dermoscopy is a useful, non-invasive, diagnostic procedure that allows us to differentiate LHS pigmented lesions from nail melanoma. Nevertheless, medication-induced melanonychia or ethnic-type melanonychia may have similar dermoscopic patterns.<sup>7</sup>

Various differential diagnoses considered for LHS can be ruled out by the appearance and etiology of hyperpigmentation. Drug-induced pigmentation usually occurs after months or years of chronic drug use, and tends to resolve once drugs are discontinued. Smoking can result in oral, mucosal pigmentation

called smoker's melanosis, and it is predominantly seen in anterior gingiva. In addition, smoking is not associated with pigmentation of the nails. Physiologic (racial) pigmentation of the oral mucosa appears in Asians, Blacks, and other dark-skinned persons, resembling LHS. Racial pigmentation is most commonly demonstrated in the gingiva although it can appear in any location.<sup>1</sup>

Laugier-Hunziker syndrome is considered a diagnosis of exclusion. Addison's disease and Peutz-Jeghers syndrome (PJS) are the main entities included in the differential diagnosis of LHS. Both are characterized by multiple oral mucosal macules. However, patients with Addison's disease also have low blood pressure, hyponatremia, hyperkalemia, hypoglycemia, and elevated blood urea nitrogen, due to their adrenocortical insufficiency. PJS is an autosomal dominant inherited condition featuring a combination of mucocutaneous pigmentation, intestinal polyposis and, importantly, predisposition to various types of cancer.<sup>6</sup> Other differential diagnoses include Bandler syndrome (BS) and McCune-Albright syndrome (MCAS) but they both emerge at a much earlier age, with BS lesions present at birth. BS is also characterized by intestinal, vascular malformation that can cause significant gastrointestinal bleeding. MCAS includes labial and genital pigmentation but

is often unilateral and does not involve the nails. The disease is also accompanied by precocious puberty in females and fibrous dysplasia.<sup>1</sup>

Several case reports have described LHS associated with esophageal melanocytosis, actinic lichen planus, hypocellular marrow and thrombocytopenia, invasive melanoma, and lupus erythematosus.<sup>8</sup> There have still not been any publications about LHS and BP. The links with these conditions have not been well established, implying that they could all be a case of coincidence.

Laugier-Hunziker syndrome is considered a chronic progressive condition, though asymptomatic. Hence, treatment for LHS is not indicated, except for esthetic and/or psychological reasons. Successful ablation of pigmented lesions has been reported using different laser devices, such as Q-switched Nd-YAG and alexandrite laser.<sup>9,10</sup> Unfortunately, recurrence levels are high, regardless of the laser technique applied.<sup>10</sup>

Laugier-Hunziker syndrome should always be considered in the differential diagnosis of middle-aged patients presenting with mucocutaneous and nail hyperpigmentation, without systemic signs or symptoms. By identifying LHS, other, more severe pigmentary diseases can be excluded and therefore other, unnecessary diagnostic procedures avoided. □

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