# Do you know this syndrome? *

## Você conhece esta Síndrome?

Priscilla Maria Rodrigues Pereira [1]
Livia Lima de Lima [3]
Adriana Valquíria de Oliveira Mariano [5]
Carlos Alberto Chirano Rodrigues [2]
Sandra Adolfina Romero Reyes [4]

**CASE REPORT**

A 64-year old woman with hyperpigmented macules on her fingernails, toenails and lips sought medical care at a dermatology outpatient clinic. The condition had been present for at least two years. Dermatological examination revealed the presence of brownish, well-defined macules in a lentiginous pattern, bilaterally located in the oral mucosa, with pigmentation similar to that of the lips and labial commissures (Figure 1). Homogenous hyperpigmentation was found in symmetrical longitudinal stripes on the nails of the 1st, 2nd and 3rd fingers, and on the nails of the 1st and 2nd toes (Figures 2 and 3). Similar pigmentation was also present on the proximal nail fold of the 2nd and 3rd fingers of the right hand, with poorly defined borders, characterizing Hutchinson's sign (Figure 4). No nail dystrophy was found and there were no macules in the conjunctiva, genital mucosa or on any other area of the body.

Histopathology of samples from the nail plate and oral mucosa showed only lentiginous hyperpigmentation of the basal layer with no cell atypia, an accumulation of melanophages and a deposit of free melanin in the superficial dermis (Figure 5). Upper and lower digestive endoscopy revealed normal mucosal epithelium with no polyps or masses. Family history was negative for pigmentary or genetic diseases. Based on the dermatological and histological findings, the diagnosis was reached and the patient was informed with respect to the benign nature of her condition and the absence of any systemic repercussions.

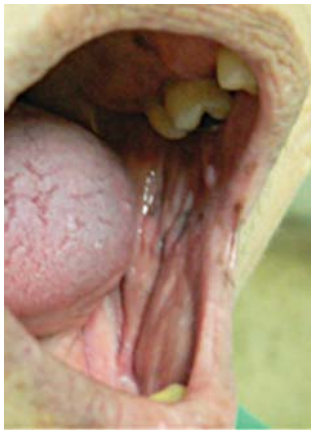

**FIGURE 1:** Hyperpigmented macules on the oral mucosa and lips, including punctiform macules in the labial commissure

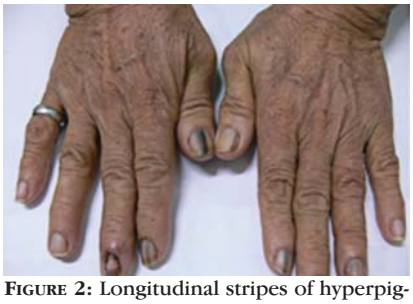

**FIGURE 2:** Longitudinal stripes of hyperpigmentation on fingernails of both hands

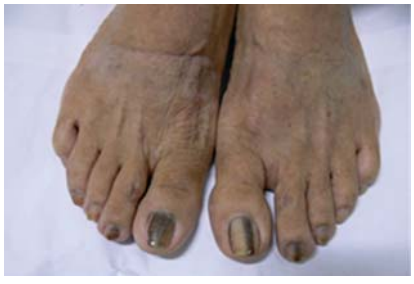

**FIGURE 3:** Longitudinal melanonychia of the toenails

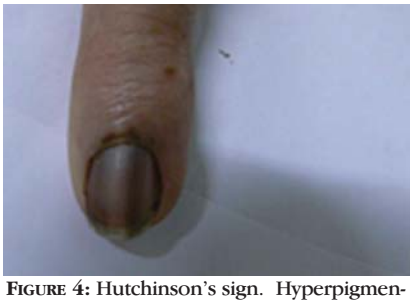

**FIGURE 4:** Hutchinson's sign. Hyperpigmentation of the proximal nail fold of the finger

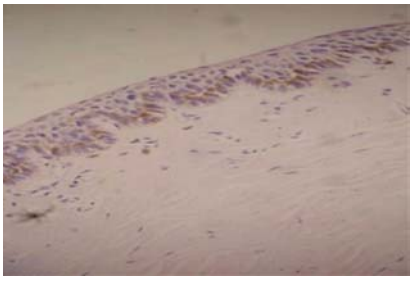

**FIGURE 5:** Lentiginous hyperpigmentation of the basal layer without cell atypia

Approved by the Editorial Board and accepted for publication on 06.04.2010.
* Work conducted in the Foundation of Tropical Dermatology and Venereologia Alfredo da Matta.
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

[1] Currently participating in the Medical Residency Program in Dermatology, Getúlio Vargas Teaching Hospital, Federal University of Amazonas, Manaus, AM, Brazil.
[2] Dermatologist, Alfredo da Matta Foundation, Manaus, AM, Brazil. Preceptor of the medical residency program, Alfredo da Matta Foundation, Manaus, AM, Brazil..
[3] Currently participating in the Medical Residency Program in Dermatology, Alfredo da Matta Foundation, Manaus, AM, Brazil. .
[4] Currently participating in the Medical Residency Program in Dermatology, Getúlio Vargas Teaching Hospital, Federal University of Amazonas, Manaus, AM, Brazil.
[5] Currently participating in the Medical Residency Program in Dermatology, Getúlio Vargas Teaching Hospital, Federal University of Amazonas, Manaus, AM, Brazil.

**WHAT SYNDROME IS THIS?**

***Laugier-Hunziker Syndrome***

Laugier-Hunziker syndrome (LHS) was first described in 1970 as melanotic pigmentation of the lips and oral mucosa that could also be accompanied by hyperpigmentation of the nails. [1] This original concept of this acquired pigmentary disorder was later modified to *idiopathic lenticular mucocutaneous pigmentation* after the condition was detected in other areas where a similar histology was found. [2,3] The etiopathogenesis of this condition is unknown, although different theories have been proposed to explain the mechanism behind the melanocytic dysfunction. [2,3]

It is most prevalent in women and in general begins between the ages of 30 and 50 years. It occurs almost exclusively in whites, unlike the patient in the current report, who is brown. [4] The syndrome is characterized by a varying number of asymptomatic, lentiginous or linear, hyperpigmented, mucocutaneous macules. [3,6] Their coloring ranges from brown to black. They may be single or confluent and generally involve the mouth and lips. [5,6] Less commonly, the syndrome may affect the labial commissure, gum, tongue, palate, fingers and toes, neck, abdomen, esophagus and the palmar and plantar regions. [3,5,6] The nails are affected in 60% of cases, although this feature has never been seen in children. When affected, one or two homogenous, longitudinal stripes may appear on the nail plate, affecting it either partially or completely. [1,5-7]

Diagnosis is clinical and was reached following the exclusion of all other causes of mucocutaneous hyperpigmentation commonly associated with longitudinal melanonychia. [1] The pathology of this condition consists of hyperpigmentation of the basal layer, an increase in melanophages and the presence of pigmentary incontinence in the papillary dermis, with no hyperproliferation or melanocytic atypia. [2,6,7] The disorder is more evident in the areas most affected. [2] Mild acanthosis has been described in some reports. [2] Therefore, it is a functional change in the melanocyte of an as yet unknown cause; however, it is probably a consequence of chronic stimulation due to enzymatic hyperreactivity in the biosynthesis of melanin-tyrosin, which subsequently results in an increase in the number of melanosomes. [2] Various conditions should be included in the differential diagnosis of mucocutaneous pigmentary disorders such as the following syndromes: McCune-Albright, LEOPARD, Addison, LAMB, Gardner, Cronkhite-Canada, neurofibromatosis and Peutz-Jeghers, as well as racial pigmentation, which is a benign disorder and the principal cause of longitudinal melanonychia. [1,6,7,8] Nail disorders may be caused by fungi such as Scytalidium dimidiatum, Fusarium spp, Wangiella dermatitidis, Exophiala dermatitidis, Scopulariopsis brevicaulis, Aspergillus niger, Alternaria sp, Penicillium sp and Acremonium sp; however, nail disorders have also been reported to have been caused by the Trichophyton rubrum dermatophyte and by yeasts (Candida spp). [8] Melanonychia may also be a consequence of the use of certain drugs, principally tetracycline, minocycline, antimalarials, phenothiazines and chemotherapeutic agents. [7] Of all the causes of localized mucosal pigmentation, the most important is amalgam tattoo, melanoma and Kaposi's sarcoma. These conditions are easily excluded by associated clinical findings and supplementary tests. [1,2,5,7,9]

Hutchinson's sign is always present in the case of subungual melanomas; however, it may be present in Peutz-Jeghers and Laugier-Hunziker syndromes, subungual hematoma, racial pigmentation and AIDS, in association with the use of certain drugs such as minocycline and zidovudine, and in Bowen's disease. [9,10]

No treatment is required, since the condition is not associated with systemic diseases or complications. [1,3,4,9] There have been no reports of malignant degeneration. [6] From a cosmetic point of view, laser therapy may be useful. [9]

Mucocutaneous hyperpigmentation is not uncommon in clinical practice and may represent a diagnosis of various different conditions. It is important to include Laugier-Hunziker syndrome in this work-up, since it constitutes a diagnosis of exclusion and its prognosis is excellent. ❑

**Abstract:** Laugier-Hunziker syndrome is an acquired macular hyperpigmentation of the oral and genital mucosa, often associated with longitudinal melanonychia. We report a case of longitudinal melanonychia on ?ngernails and toenails, hutchinson's signal and lenticular pigmentation of her lips and oral mucosa. Histopathological analysis is tipical and had a broad differential diagnosis. This is a rare and benign entity, which should be included in the differential diagnosis of mucocutaneous hyperpigmentation.
Keywords: Diagnosis, differential; Mouth mucosa; Pigmentation disorders

**Resumo:** Síndrome de Laugier-Hunziker é caracterizada por hiperpigmentação macular adquirida, idiopática das mucosas oral e genital, que pode estar associada à melanoníquia longitudinal. Relata-se caso de melanoníquia longitudinal das mãos e pés, sinal de Hutchinson e pigmentação lenticular dos lábios e mucosa oral. A histologia é típica, com amplo diagnóstico diferencial. Trata-se de entidade rara e benigna, que deve ser incluída na diagnose diferencial das hiperpigmentações mucocutâneas.
Palavras-chave: Diagnóstico diferencial; Hiperpigmentação; Mucosa bucal

**REFERENCES**

1. Gencoglan G, Gerceker-Turk B, Kilinc-Karaarslan I, Akalin T, Ozdemir F. Dermoscopic Findings in Laugier-Hunziker Syndrome. Arch Dermatol. 2007;143:631-3.
2. Vega Gutiérrez J, Miranda Romero A, Martínez G, Torrero MV, López de Juan M. Hyperpigmentation mimicking Laugier syndrome, levodopa therapy and Addison's disease. J Eur Acad Dermatol Venereol. 2003;17:324-7.
3. Ajith C, Handa S. Laugier-Hunziker pigmentation. Indian J Dermatol Venereol Leprol. 2005;71:354-6.
4. Tan J, Greaves MW, Lee LH. Laugier-Hunziker syndrome and hypocellular marrow: a fortuitous association? Clin Exp Dermatol. 2007;32:584-5.
5. Sardana K, Mishra D, Garg V. Laugier-Hunziker Syndrome. Indian Pediatrics. 2006;43:998-1000.
6. Aytekin S, Alp S. Laugier-Hunziker syndrome associated with actinic lichen planus. J Eur Acad Dermatol Venereol. 2004;18:221-42.
7. Lampe AK, Hampton PJ, Woodford-Richens K, Tomlinson I, Lawrence CM, Douglas FS. Laugier-Hunziker syndrome: an important differential diagnosis for Peutz-Jeghers syndrome. J Med Genet. 2003;40:77.
8. Miot HA, Gumieiro JH, Poli JPV, Camargo RMP. Caso para diagnóstico. Melanoníquia fúngica. An Bras Dermatol. 2007;82:480-2
9. Ayoub N, Barete S, Bouaziz JD, Le Pelletier F, Frances C. Additional conjunctival and penile pigmentation in Laugier-Hunziker syndrome: a report of two cases. Int J Dermatol. 2004;43:571-4.
10. Mendonça IRS, Kac BK, Silva RT, Spinelli LP, Orofino RR, Andrade JRF. Melanoma do aparelho ungueal. An Bras Dermatol. 2004;79:575-580.

*MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:*
*Priscilla Maria Rodrigues Pereira*
*Avenida Codajás, 24 Cachoeirinha Manaus – (AM), Brazil.*
*Telephone.: +55 92 3663 4747*

How to cite this article/*Como citar este artigo*: Pereira PMR, Rodrigues CAC, Lima LL, Reyes SAR, Mariano AVO. Do you know this syndrome? Laugier-Hunziker syndrome. An Bras Dermatol. 2010;85(5):751-3.