A 58-year-old female with a medical history of type 2 diabetes mellitus and arterial hypertension, presented with 3 months history of multiple hyperpigmented macules in the labial mucosa, hard palate, and tongue without any associated symptoms [Figure 1a and b]. The patient was a nonsmoker and denied the use of any new medication that could cause the pigmentation. Dermatological examination revealed longitudinal melanonychia on the first toenail of her right foot [Figure 2]. The patient denied the history of abdominal pain, bleeding, diarrhea, intussusception, or intestinal obstruction. The family history of gastrointestinal neoplasia or pigmentary disorders was negative. Laboratory tests revealed elevated blood glucose (206 mg/dL) and urinary glucose (250 mg/dL). Complete blood count was unremarkable, fecal immunochemical test resulted negative, and levels of adrenocorticotropic hormone and cortisol were all within normal limits. Labial mucosa biopsy revealed an epidermis with acanthosis and pigmentation of the basal layer [Figure 3]. Potassium hydroxide examination of the nail was negative for the presence of hyphae, pseudohyphae, or yeast cells.

**Question**

What is your diagnosis?

**Figure 1:** (a and b) Presence of multiple hyperpigmented macules in labial mucosa, hard palate and tongue

**Figure 2:** Longitudinal melanonychia in first right foot toe nail

**Figure 3:** Histopathological findings: Epidermis with acanthosis and pigmentation of the basal layer (H and E, ×100)
Answer
Laugier-Hunziker syndrome (LHS).

Diagnosis
Due to the clinical findings of multiple hyperpigmented macules in oral and labial mucosa associated with longitudinal melanonychia in the absence of systemic disease, the patient was diagnosed with LHS.

Review
LHS is a rare, benign and acquired pigmentary disorder first described in 1970 by Laugier and Hunziker; characterized by the presence of multiple hyperpigmented macules in oral and labial mucosa. In 1979, Baran emphasized the importance of nail involvement as an essential finding for its diagnosis, which is found in 60% of cases. Reported nail abnormalities include a homogenous involvement of one half of the nail, an isolated pigmented band and double striking of the nails. This syndrome of unknown etiology, usually affects Caucasian middle-aged women, and has not been associated with any systemic diseases.

It is a diagnosis of exclusion and a thorough examination is warranted to rule out other pathologies. Differential diagnoses of this entity include Peutz-Jeghers syndrome (PJS), Addison’s disease, Albright syndrome, Bandler syndrome, Cronkhite-Canada syndrome, melanoma, drug induced pigmentation (phenytoin, zidovudine, antimalarials, clofazimine and phenothiazine), hiperpigmentation due to smoking or heavy metal exposure and oral lichen planus.

Typically histopathologic examination shows hyperpigmentation of the basal epithelial layer. Most reports suggest melanocytes are normal in number, morphology and distribution. In some cases melanophages in the papillary dermis may be observed. These findings indicate that the condition is due to an increased melanocytic activity. No putative genetic defects have been identified, and this is generally thought to be an acquired condition; however, STK11 gene testing allows clinicians to differentiate between PJS and LHS.

LHS is a benign condition and no treatment is needed. However, some patients may choose to remove the hyperpigmentation for aesthetic considerations. Cryotherapy, Q-switched Nd:YAG laser and Q-switched alexandrite laser have been successfully used to remove the hyperpigmented macules.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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