Foresee Your Next Patient

Laugier-Hunziker Syndrome

Brooke Evans, MPH[1] • Richard Byrd, MD[1,2]

During a routine wellness examination, an otherwise healthy 12-year-old boy of Indian descent had concerns about dark areas on his tongue. He had first noticed them 1 year ago, and they had gradually increased in number and size.

He denied any associated discomfort, and results of a review of systems were negative for fatigue, weight loss, weakness, abdominal pain, vomiting, rectal bleeding, anemia, drug exposure, bone pain, or precocious puberty. His family history was positive for similar spots on his mother's tongue (findings of a biopsy had been benign). There was no family history of intestinal polyps or gastrointestinal (GI) tract malignancy. He denied taking any long-term medications.

Physical examination findings were significant for several hyperpigmented macules measuring 1 to 5 mm over the dorsolateral tongue, hyperpigmentation along the maxillary and mandibular gingiva, a 1-cm area of gray-brown pigmentation on the buccal mucosa at the opening of the parotid duct bilaterally, and longitudinal hyperpigmented bands on the nails of both index fingers and the left second toe (**Figures 1-6**). His lips and genitalia were without altered pigmentation.

Given the patient's oral and ungual hyperpigmentation with onset at puberty, gradual progression, absence of systemic symptoms, and negative family history of intestinal polyps or GI tract malignancy, a diagnosis of Laugier-Hunziker syndrome (LHS) was made. The patient and his mother were counseled regarding its benign nature.

Discussion. LHS, also known as idiopathic lenticular mucocutaneous pigmentation, is a benign pigmentary disorder without systemic symptoms. It is associated with lenticular or linear hyperpigmented macules that can be isolated or confluent and that can present in various mucocutaneous tissues. It can be either familial or sporadic, with the latter being most common.[1,2] The average age of onset is 52 years, and the condition most commonly occurs in white adults.[3] A higher incidence in females has been described.[4]

The most common sites of involvement are the mouth (especially the buccal mucosa) and lips. Less frequent locations include the corners of the mouth, gingivae, tongue, fingers, and plantar aspect of feet.[5] Nail lesions are present in about 60% of adult cases, and such findings include pigmented bands of the nail plate (longitudinal melanonychia). This finding, however, is far less common in the pediatric population.[6] In the absence of systemic symptoms or specific drug or heavy metal exposure, the presence of both oral and nail pigmentation is highly suggestive of LHS.[7]

Laugier-Hunziker syndrome is associated with lenticular or linear hyperpigmented macules that can be isolated or confluent and that can present in various mucocutaneous tissues.

A biopsy of lesions, although not necessary, may help to confirm the diagnosis. Histopathologic findings include increased pigmentation of the basal layer with normal quantity and morphology of melanocytes, epidermal basement membrane pigmentation, and pigment-laden macrophages in the papillary dermis.[3,8] Dermoscopic findings are tissue-specific: A parallel banded pattern is found in lesions of the lips and genitalia, and a single homogenous longitudinal band of pigmentation is shown in nail lesions.[9]

LHS is a clinical diagnosis of exclusion. Other conditions such as Peutz-Jeghers syndrome, Addison disease, subungual melanoma, drug-related hyperpigmentation, heavy metal exposure, and benign racial hyperpigmentation should be considered (**Table**).[3,5,7,10-13]

AFFILIATIONS:
[1]Texas A&M College of Medicine, Houston, Texas
[2]Kelsey-Seybold Clinic, Houston, Texas

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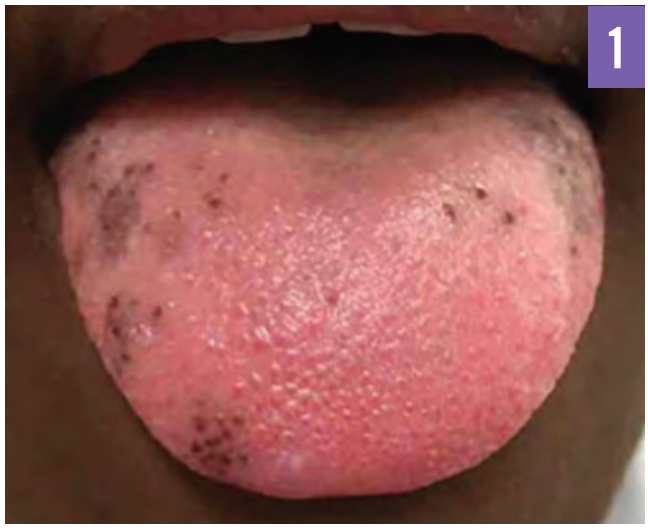
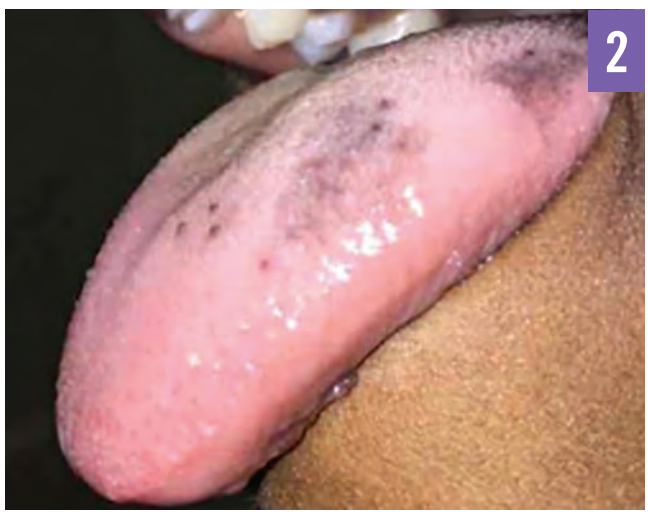
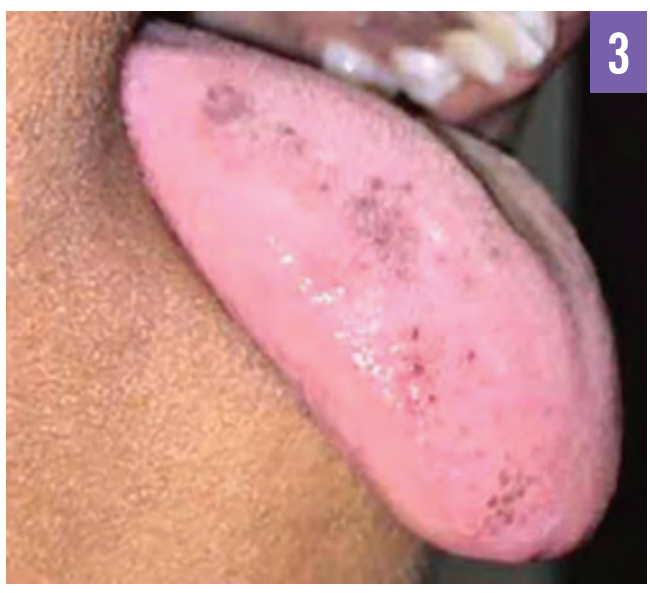
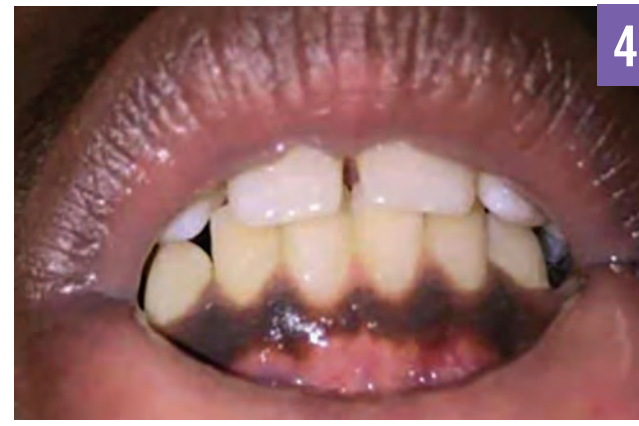
CORRESPONDENCE:
Richard Byrd, MD, Kelsey-Seybold Clinic, 7010 Hwy 6, Missouri City, TX 77459 (richard.byrd@kelsey-seybold.com)



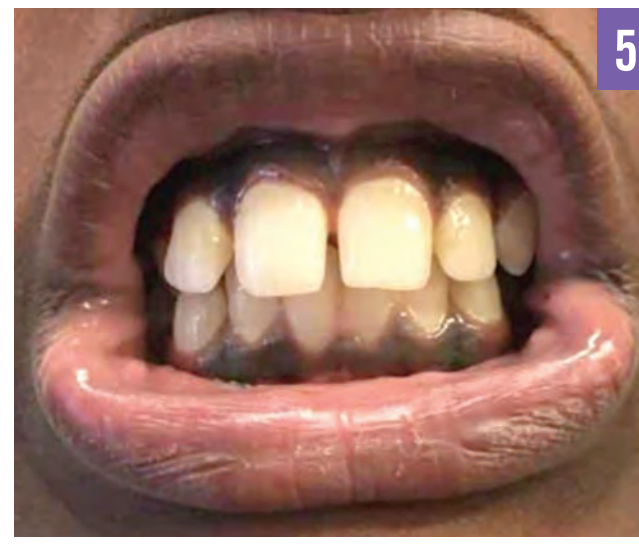




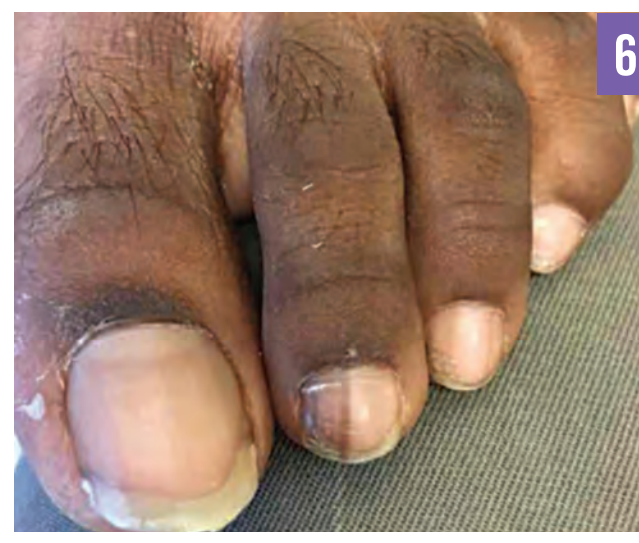
Bilateral hyperpigmentation of the patient's tongue.



Mandibular gingival hyperpigmentation.



Maxillary and mandibular gingival hyperpigmentation.



Longitudinal melanonychia on the patient's left second toe.

Table. Differential Diagnosis in Children With Mucocutaneous Hyperpigmentation[3,5,7,10-13]

Diagnosis	Etiology	Hyperpigmentation Characteristics	Systemic Manifestations
Laugier-Hunziker syndrome	Acquired benign mucocutaneous hyperpigmentation	Mouth (buccal mucosa, labial pigmentation confined to lips), nails; onset at puberty or adulthood (average age of onset, 52 y)[3]	Benign without systemic manifestations
Peutz-Jeghers syndrome	Autosomal dominant mutation of serine/threonine kinase 11 gene (*STK11*)[13]	Mouth, nostrils, hands, feet; rarely involve nails; onset at birth or infancy[7]	Associated with intestinal polyposis and malignancies of the colon, pancreas, small intestine, stomach, breast, ovaries, and lung[7]
Addison disease	Acquired or congenital primary adrenal insufficiency	Diffuse, mostly on areas subject to sun exposure, trauma, or friction (face, skin creases)[5]; oral hyperpigmentation is an early sign of disease	Often presents with abdominal pain, weakness, fatigue, weight loss
Subungual melanoma	Acquired	Brown to black nail band with irregular border, rapid increase in size, extension of pigmentation to proximal or lateral nail folds (Hutchinson sign)[5]; extremely rare in children[11,12]	Malignant proliferation of cells; highly metastatic with poor prognosis
Drug-related hyperpigmentation	Specific exposure to drugs such as antimalarials, chemotherapeutics, and oral contraceptives[10]	Diffuse oral and nail lesions in the setting of long-term use of associated drug[10]; often resolves after discontinuation of offending agent[5]	None
Heavy metal exposure	Specific exposure	Varied presentation of oral pigmentation in the setting of known or suspected heavy metal exposure (eg, blue-black discoloration of gingival margin with lead; gray with bismuth and mercury)[13]	Variable; associated with specific heavy metal toxicities
Racial hyperpigmentation	Acquired or congenital hyperpigmentation in nonwhite individuals	Most often gingival but can occur elsewhere[5]	None

Because of the nonsystemic and benign nature of LHS, no treatment is indicated. However, patients may seek therapies for cosmetic purposes. Treatments using laser therapy and cryotherapy have been used to reduce areas of skin hyperpigmentation.[10]

Our patient, a 12-year-old boy of Indian descent, represents a patient population that is not well represented in the LHS literature. At a follow-up appointment 4 months later, his pigmentation remained unchanged. The family was again reassured of the syndrome's benign nature. ■

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