Hyperpigmented Papules on the Tongue of a Child

Helena Drolshagen, MD; Blake Chandler, MD; Megan S. Evans, MD

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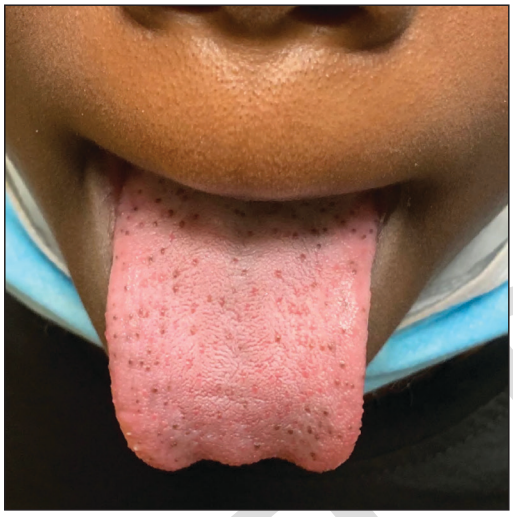

A 9-year-old Black boy presented to the dermatology clinic for evaluation of dark spots on the tongue. The family first noted these spots 5 months prior and reported that they remained stable during that time. The patient's medical history was notable for autism spectrum disorder and multiple food allergies. His family history was negative for similar oral pigmentation or other pigmentary anomalies. A review of systems was positive only for selective eating and rare nosebleeds. Physical examination revealed numerous dark brown, pinpoint papules across the dorsal aspect of the tongue. No hyperpigmentation of the buccal mucosae, lips, palms, or soles was identified. Several light brown streaks were present on the fingernails and toenails, consistent with longitudinal melanonychia. A prior complete blood cell count was within reference range.

WHAT'S YOUR **DIAGNOSIS?**

a. Addison disease
b. black hairy tongue (or lingua villosa nigra)
c. hereditary hemorrhagic telangiectasia
d. Peutz-Jeghers syndrome
e. pigmented fungiform papillae of the tongue

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Drs. Drolshagen and Chandler are from the Department of Internal Medicine, Baptist Health–University of Arkansas for Medical Sciences, North Little Rock. Dr. Evans is from the Department of Dermatology, University of Arkansas for Medical Sciences, Little Rock.

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Correspondence: Helena Drolshagen, MD, 3201 Springhill Dr, North Little Rock, AR 72117 (helena.drolshagen@baptist-health.org).

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THE **DIAGNOSIS:** Pigmented Fungiform Papillae of the Tongue

Our patient's hyperpigmentation was confined to the fungiform papillae, leading to a diagnosis of pigmented fungiform papillae of the tongue (PFPT). A biopsy was not performed, and reassurance was provided regarding the benign nature of this finding, which did not require treatment.

Pigmented fungiform papillae of the tongue is a benign, nonprogressive, asymptomatic pigmentary condition that is most common among patients with skin of color and typically develops within the second or third decade of life.[1,2] The pathogenesis is unclear, but activation of subepithelial melanophages without evidence of inflammation has been implicated.[2] Although no standard treatment exists, cosmetic improvement with the use of the Q-switched ruby laser has been reported.[3,4] Clinically, PFPT presents as asymptomatic hyperpigmentation confined to the fungiform papillae along the anterior and lateral portions of the tongue.[1,2] Pigmented fungiform papillae of the tongue typically is an isolated finding but rarely can be associated with hyperpigmentation of the nails (as in our patient) or gingiva.[2] Three different clinical patterns of presentation have been described: (1) a single well-circumscribed collection of pigmented fungiform papillae, (2) few scattered pigmented fungiform papillae admixed with many nonpigmented fungiform papillae, or (3) pigmentation of all fungiform papillae on the dorsal aspect of the tongue.[2,5,6]

Pigmented fungiform papillae of the tongue is a clinical diagnosis based on visual recognition. Dermoscopic examination revealing a cobblestonelike or rose petal–like pattern may be helpful in diagnosing PFPT.[2,5-7] Although not typically recommended in the evaluation of PFPT, a biopsy will reveal papillary structures with hyperpigmentation of basilar keratinocytes as well as melanophages in the lamina propria.[8] The latter finding suggests a transient inflammatory process despite the hallmark absence of inflammation.[5] Melanocytic neoplasia and exogenous granules of pigment typically are not seen.[8]

Other conditions that may present with dark-colored macules or papules on the tongue should be considered in the evaluation of a patient with these clinical findings. Black hairy tongue (BHT), or lingua villosa nigra, is a benign finding due to filiform papillae hypertrophy on the dorsum of the tongue.[9] Food particle debris caught in BHT can lead to porphyrin production by chromogenic bacteria and fungi. These porphyrins result in discoloration ranging from brown-black to yellow and green occurring anteriorly to the circumvallate papillae while usually sparing the tip and lateral sides of the tongue. Dermoscopy can show thin discolored fibers with a hairy appearance. Although normal filiform papillae are less than 1-mm long, 3-mm long papillae are considered diagnostic of BHT.[9] Treatment includes effective oral hygiene and desquamation measures, which can lead to complete resolution.[10]

Peutz-Jeghers syndrome is a rare genodermatosis that is characterized by focal hyperpigmentation and multiple gastrointestinal mucosal hamartomatous polyps. Peutz-Jeghers syndrome should be suspected in a patient with discrete, 1- to 5-mm, brown to black macules on the perioral or periocular skin, tongue, genitals, palms, soles, and buccal mucosa with a history of abdominal symptoms.[11,12]

Addison disease, or primary adrenal insufficiency, may present with brown hyperpigmentation on chronically sun-exposed areas; regions of friction or pressure; surrounding scar tissue; and mucosal surfaces such as the tongue, inner surface of the lip, and buccal and gingival mucosa.[13] Addison disease is differentiated from PFPT by a more generalized hyperpigmentation due to increased melanin production as well as the presence of systemic symptoms related to hypocortisolism. The pigmentation seen on the buccal mucosa in Addison disease is patchy and diffuse, and histology reveals basal melanin hyperpigmentation with superficial dermal melanophages.[13]

Hereditary hemorrhagic telangiectasia is an inherited disorder featuring telangiectasia and generally appears in the third decade of life.[14] Telangiectases classically are 1 to 3 mm in diameter with or without slight elevation. Dermoscopic findings include small red clots, lacunae, and serpentine or linear vessels arranged in a radial conformation surrounding a homogenous pink center.[15] These telangiectases typically occur on the skin or mucosa, particularly the face, lips, tongue, nail beds, and nasal mucosa; however, any organ can be affected with arteriovenous malformations. Recurrent epistaxis occurs in more than half of patients with hereditary hemorrhagic telangiectasia.[14] Histopathology reveals dilated vessels and lacunae near the dermoepidermal junction displacing the epidermis and papillary dermis.[15] It is distinguished from PFPT by the vascular nature of the lesions and by the presence of other characteristic symptoms such as recurrent epistaxis and visceral arteriovenous malformations.

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