Hyperpigmented macules in the oral mucosa and melanonychia

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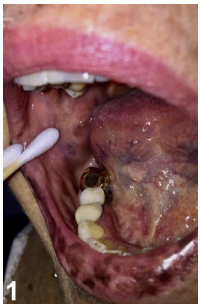

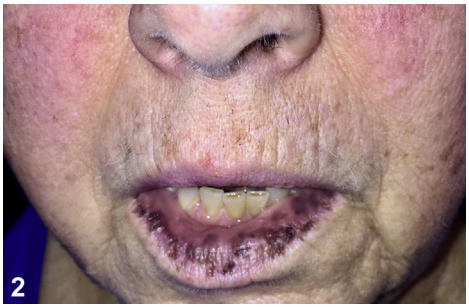

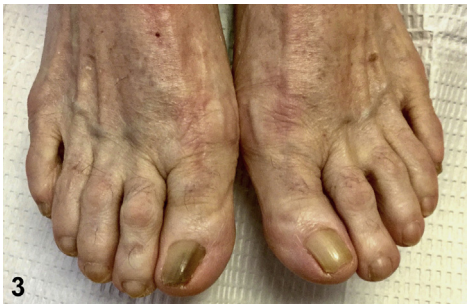

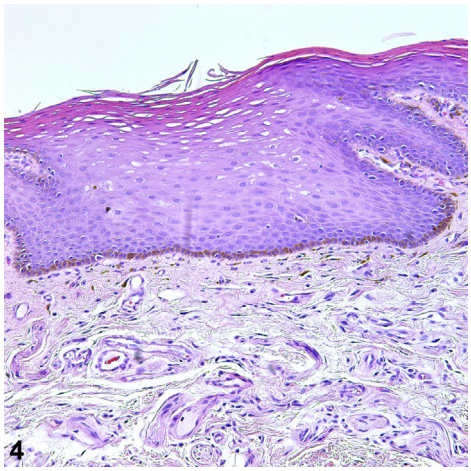

CASE PRESENTATION

A 69-year-old woman presented with a 15-year history of asymptomatic, dark spots gradually increasing in number on the lips and oral mucosa. Physical examination found coalescing, hyperpigmented macules on the buccal mucosa, palate, and lower lip mucosa (Figs 1 and 2). Longitudinal melanonychia was noted on the right great toe (Fig 3). Medical history was unremarkable. Social history was significant for a 40 pack-year smoking history. All age-appropriate routine screenings were up to date and unremarkable. Family history was negative for oral pigmentation, colon polyps, and cancer. A punch biopsy specimen was taken from the buccal cheek (Fig 4).

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Question 1: What is the most likely diagnosis?

A. Peutz-Jeghers syndrome

B. Laugier-Hunziker syndrome

C. Addison disease

D. Smoker's melanosis

E. LAMB Syndrome

Answers:

A. Peutz-Jeghers syndrome — Incorrect. Although Peutz-Jeghers syndrome has a similar presentation, the patient lacked a family history of the disease, gastrointestinal polyps, and pigmented macules in infancy or early childhood. Additionally, Peutz-Jeghers syndrome typically spares the tongue, palate, and fingernails.

B. Laugier-Hunziker syndrome — Correct. Laugier-Hunziker syndrome is a benign, acquired condition that involves gradual progression of asymptomatic macular hyperpigmentation of the mucocutaneous surfaces and nails.

C. Addison disease — Incorrect. Hyperpigmentation in Addison disease is found in areas subject to pressure.[1] In addition, this patient did not exhibit any signs of insufficient cortisol or aldosterone production.

D. Smoker's melanosis — Incorrect. Smoker's melanosis presents with diffuse patches or numerous small melanotic macules in the anterior labial gingiva of the mandible.[2] The oral distribution and nail involvement makes smoker's melanosis unlikely.

E. LAMB Syndrome — Incorrect. LAMB syndrome, now referred to as *Carney complex*, is a multiple endocrine neoplasia syndrome characterized by lentigines, atrial myxomas, and blue nevi. Lentigines in LAMB syndrome typically manifest in the first 2 decades of life, and there is no mention of blue nevi or cardiac findings in this patient's history.

Question 2: What is the most common extraoral site of pigmentation in this condition?

A. Palms and soles

B. Nails

C. Sclera

D. Anogenital mucosa

E. Small bowel

Answers:

A. Palms and soles — Incorrect. There have been few reported cases involving the palms and soles. Addison's disease is characterized by pronounced hyperpigmentation of the palms and soles in addition to the perineum, axillae, and areaolae.[3]

B. Nails — Correct. Melanonychia has been described in up to 60% of cases of Laugier-Hunziker syndrome.[4] Nail involvement usually presents as single- or double-striped longitudinal lines. Fingernails are more commonly affected than toenails.

C. Sclera— Incorrect. Cases involving hyperpigmentation of the sclera and conjunctiva have rarely been reported. Nail pigmentation has been reported in more than half of all Laugier-Hunziker syndrome cases.

D. Anogenital mucosa — Incorrect. Cases involving hyperpigmentation of the anogenital mucosa have been reported, although these findings occur less frequently than nail involvement.

E. Small bowel — Incorrect. Small bowel pigmentation is not associated with Laugier-Hunziker syndrome. However, in Peutz-Jeghers syndrome, polyps have a predilection for the small bowel.

Question 3: What is the most effective treatment for this condition?

A. Glucocorticoid and mineralocorticoid therapy

B. Smoking cessation

C. Psoralen plus ultraviolet A (PUVA) therapy

D. No treatment necessary

E. Hydroquinone

Answers:

A. Glucocorticoid and mineralocorticoid therapy — Incorrect. Long-term glucocorticoid and mineralocorticoid therapy is effective for hyperpigmentation in Addison disease, which is a primary adrenocortical insufficiency disorder.

B. Smoking cessation — Incorrect. Although smoking cessation is beneficial for the patient's overall health, Laugier-Hunziker syndrome is not associated with smoking. Hyperpigmentation caused by smoker's melanosis is likely to resolve spontaneously after several years of smoking discontinuation.[2]

C. PUVA therapy — PUVA radiation treatment is used for psoriasis, mycosis fungoides, eczema, and vitiligo,

but not Laugier-Hunziker syndrome. Contrarily, lentigines are a well-recognized complication of PUVA therapy.[5]

D. No treatment necessary — Correct. Laugier-Hunziker syndrome is a benign condition that does not require treatment other than for cosmetic reasons. Cosmetic treatment modalities that have been successfully used include cryosurgery and ablation with Q-switched alexandrite, neodymium-doped yttrium aluminium garnet (Nd:YAG), and diode lasers.[1]

E. Hydroquinone — Incorrect. Hydroquinone is a topical skin-lightening agent that should be avoided in the eyes and mucosa. More effective cosmetic treatment options for disfiguring or bothersome lesions include cryotherapy and laser therapy.

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