Laugier-Hunziker Syndrome

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Laugier-Hunziker syndrome (LHS) is a rare acquired benign hyperpigmentation of the oral mucosa and lips which is often associated with longitudinal melanonychia and spotted macular pigmentation of the acral skin. We herein report two patients exhibiting the typical features of LHS. (Dermatol Sinica 24: 209-212, 2006)

Key words: Laugier-Hunziker Syndrome, Longitudinal melanonychia

INTRODUCTION

Laugier-Hunziker syndrome (LHS) was first described in 1970.¹ It is characterized by asymptomatic hyperpigmented macules of the oral mucosa, lips and acral skin and pigmented longitudinal bands of the nails. The onset of the condition is usually in early or mid adult life.² We herein report two typical cases of LHS.

CASE REPORT

Case 1

An 84-year-old Chinese female presented with a 40-year history of hyperpigmentation on the oral mucosa, lips, hands, feet, and nails. There was no family history of mucocutaneous pigmentation or intestinal polyps. Physical examination revealed many variously sized round, oval or irregularly shaped brown macules were on the conjunctivae, lips, tongue, palate, buccal and gingival mucosa (Fig. 1A), and were also found on the distal parts of the hands and feet (Fig. 1B and 1C). Besides, 1-2 mm wide, brown longitudinal streaks were noted on some fingernails and toenails (Fig. 1D).
No periorificial lesion was noted.

Results of routine laboratory tests were all within the normal range, as the serum assays for thyrotropin, T3, T4, adrenotropin, and cortisol. Over the past 30 years she had had several examinations of x-ray studies of barium enema and upper GI series, oesophagastroduodenoscopies and colonoscopies which disclosed no polyp. A previous histological examination of the skin lesions at another hospital showed acanthosis and basal layer hyperpigmentation.

Case 2

A 72-year-old Chinese female had a 30-year history of hyperpigmentation on the oral mucosa, lips, and right thumb. She also noticed vertical brown lines running along three fingers in recent 2 years. There was no family history of mucocutaneous pigmentation or intestinal polyps. Dermatological examination revealed many variously sized and shaped brown macules on the lips, tongue, palate, and buccal and gingival mucosa (Fig. 2A), as well as the distal parts of the right thumb (Fig. 2B). Besides, 1-2 mm wide, brown longitudinal streaks were noted on three fingernails (Fig. 2C). No periorificial lesion was noted.

Laboratory investigations, including urine cortisol and serum thyrotropin, T3, T4, and adrenotropin were all within the normal range. Colonoscopy disclosed no hamartomatous polyposis. Incisional biopsy of the lower lip revealed basal cell layer hyperpigmentation and pigment incontinence (Fig. 3).

DISCUSSION

In 1970, Laugier and Hunziker reported five cases of acquired hyperpigmentation of the mouth and lips. Two of these patients also displayed longitudinal pigmented streaks on the
nails. The cause of this disorder, named Laugier-Hunziker syndrome (LHS), is unknown yet. LHS is a rare acquired disorder which is known to be an entirely benign condition without systemic manifestations. Most reported cases happened to white European. Initially a female predominance was suggested, but it is now thought that male and female are equally affected. Almost all known cases to date have been sporadic. However, Makhoul et al. reported the unusual occurrence of three cases of LHS in the same family.

LHS is characterized by the presence of a variable number of asymptomatic pigmented lenticular mucocutaneous macules. The colors of the lesions vary from slate to brown-black. They can be isolated or confluent. The buccal mucosa and lips are most commonly involved. The corners of the mouth, the gingivae, tongue, fingers, and the plantar aspect of the feet are less frequent locations. Rarely, pigmented macules occur on genitalia, perineal or perianal areas, sclerae and esophagus. The characteristic changes in nails are longitudinal streaks of pigment known as longitudinal melanonychia. The nail change can be found in approximately 60% of patients. The cause of nail pigmentation is unknown, but it is believed to be similar to that of the lip and mucosal pigmentation.

Histologically, there is increased basal keratinocyte melanin without expansion of the melanocytic population and superficial pigmentary incontinence with dermal melanophages. Mild to moderate acanthosis can also be noted. Electron microscopy reveals numerous mature, isolated or grouped melanosomes of variable sizes within the cytoplasm of keratinocytes and melanophages in the papillary dermis.
Several conditions must be considered in the differential diagnosis of nail and mucocutaneous pigmentary abnormalities. Such as adrenal insufficiency and nail changes resulting from administration of systemic agents, especially after chemotherapy. In our cases, Addison’s disease was eliminated on the clinical and hormonal basis. Drug ingestion did not appear relevant since the patients denied any previous or current drug intake. Peutz-Jeghers syndrome (PJS) must also be considered in the differential diagnosis. PJS is an autosomal dominant condition, in which the mucocutaneous lesions appear around birth or at an early age with mainly a periorificial distribution. Other pigmented spots can also be found on the dorsal and volar aspects of the hands and feet. There is a strong association with hamartomatous polyposis of the gastrointestinal tract and an increased risk for neoplasia. In our cases, the late onset of the disease, absence of a family history and GI polyposis make the diagnosis of PJS very unlikely. On the contrary, in addition to the aforementioned findings, the distribution of the lesions, i.e., lesions confined to the oral mucosa, lips and acral skin with sparing of the periorificial sites, and the presence of nail pigmentation are also in favor of the diagnosis of LHS.

As for our knowledge, LHS has not been reported in Taiwan. The importance of recognizing this acquired benign disorder is to avoid unnecessary and potentially hazardous investigations. No treatment is required other than reassurance.

REFERENCES