Laugier-Hunziker syndrome in a young girl

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SUMMARY

Laugier-Hunziker syndrome (LHS) is a rare, acquired disorder characterised by circumscribed pigmented macules of the oral mucosa. This rare condition usually occurs on the lips and the acral glabrous skin (mainly fingers and toes). There have been a few instances where fingernail and toenail melanonychia coexist, but not in this patient. Since very similar mucocutaneous lesions are also seen in malignancy syndromes, LHS is an exclusion diagnosis. It occurs mainly in middle-aged individuals with a ratio of approximately of 2:1 between female to male. Nonetheless, cases among children and adolescents were also reported. We describe a case report of a young Malay girl who was incidentally found with multiple oral labial lesions while presented to the clinic with other concerns.

INTRODUCTION

Laugier-Hunziker syndrome (LHS) is often diagnosed by exclusion after ruling out all other potential causes of oral and labial hyperpigmentation, such as physiologic pigmentation and inherited lentiginosis-related disorders like Peutz-Jeghers syndrome (PJS). Drug-induced pigmentation, Addison's disease (AD) and other illnesses characterised by diffuse oral mucosal pigmentation must also be considered in the differential diagnosis.

The diagnosis of LHS is supported by the absence of systemic symptoms as well as by negative results from the relevant diagnostic test.

This syndrome typically affects women of middle-aged and primarily Caucasian origin.[2] Recognition of this syndrome is essential in middle-aged patients because the development of new areas of pigmentation, especially in mucosal surfaces, may be related to malignancy. Although mucosal melanomas are uncommon, they should always be considered when making a differential diagnosis among Caucasians. Among young patients, however, there have been reports of LHS cases3 including reports on familial cases of LHS.[1]

CASE PRESENTATION

A 12-year-old Malay girl was brought by her parents to a primary care clinic in Kuantan. The parents were concerned with her noticeable rapid growth for the last year which was concurrent with her attainment of menarche. She is the youngest of four siblings and at her current age, she is the tallest compared to her elder sisters. Anthropometry examination revealed both her weight and height for age were at the 90th centile and appropriately following the mid-parental height growth chart.

There was no history of malaise, fatigue, fainting episodes, weight loss, recurrent abdominal pain, intermittent vomiting, gastrointestinal bleeding, palpitation or shortness of breath. There was no relevant drug history. There was no history of gastroesophageal reflux disease and gastritis.

Parents were non-consanguineous. There was no history of diabetes mellitus, thyroid disease, intestinal polyposis or mucocutaneous pigmentary diseases in the family. Psychosocial screening using the HEADSSS approach revealed no significant problem, especially in the dietary, sexual and mental aspects.

Her dental history was unremarkable with the last visit to a dentist being three years ago.

Upon physical examination, she appeared relaxed and comfortable in a euthyroid state. Her vital signs include a temperature of 36.7°C, a blood pressure of 100/60 mm Hg, a heart rate of 82 beats/min and a respiration rate of 14 breaths/min. There were no dysmorphic facial features or neurocognitive stigmata. The further assessment noted the patient was in Tanner stage 2.

Examination of the oral mucosa revealed multiple melanotic macules with clearly defined borders on the lower and upper labial mucosal area as shown in Figure 1. There were no similar lesions over the vermilion border and the buccal mucosa on both sides. The other mucosal surfaces, such as the oropharynx, conjunctiva, oesophagus, anus, vulva and perineum were not affected. She has no skin and fingernail or toenail involvement.

Upon incidental discovery of the oral melanotic macules, the parents admitted that they had never realised about the presence of mucosa pigmentation. The lesions were asymptomatic and the related inquiries were negative. Nevertheless, they were concerned about the lesions' progression and underlying cause.

Complete blood cell count, serum electrolytes, liver function test and chest radiograph revealed no abnormality. Referrals to paediatrician and dermatologist were made and the following investigations; serum cortisol, TSH, FSH and LH, all within normal range, and abdominal ultrasound was unremarkable.

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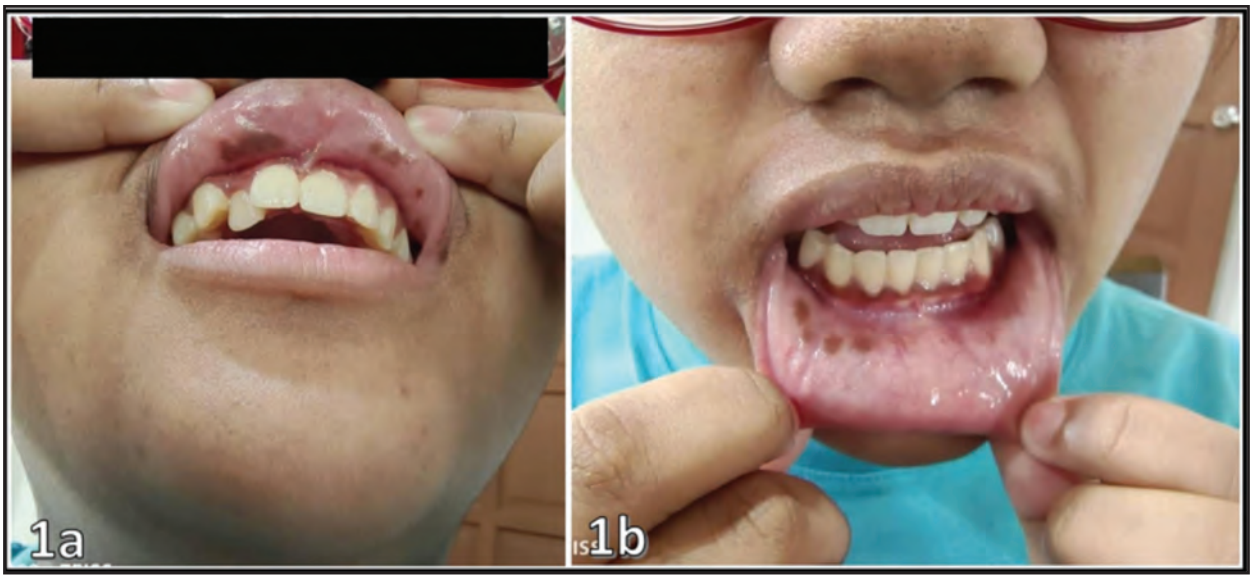

Fig. 1: (a) Hyperpigmented macules over upper labial mucosa and (b) lower labial mucosa

Given the pigmented macules on the upper and lower labial lip, no history of medication intake, and standard laboratory investigations, a diagnosis of LHS was made. The parents were reassured of the benign nature of the disorder and that treatment was unnecessary. Our patient and his parents did not have any cosmetic concerns. As such, no treatment was given. Further follow-up 6 months later noted no progression of the lesions with no psychosocial concern identified.

DISCUSSION

LHS is an uncommon condition with few reported cases. Previous research revealed a 2:1 overall female-to-male ratio, indicating a predominance of women. After carefully going over the information, a preference for women became evident. There were 76 patients in total, and 55 (72.37%) were women.[4] The illness typically affects people between the ages of 40 and 55, with a mean age of onset of 50 years, per study.[4,6] It can sporadically happen before puberty as in this patient.[6,7]

The oral lesion in LHS can be lenticular, circular, linear in shape, single or confluent, brown, black, or grey, with a smooth surface and clearly or vaguely defined edges. Similar lesions may also exist in other mucosal surfaces, such as the oropharynx, conjunctiva, oesophagus, anus, vulva and perineum, as well as the facial skin, belly and other places.[4] Up to 60% of instances can affect nails, but in this patient, there was no evidence of fingernail or toenail melanonychia.[1] By screening out other conditions that can also lead to high mucocutaneous pigmentation, LHS is diagnosed. The differential diagnosis should include PJS and AD.

PJS is an uncommon form of autosomal dominant genodermatosis that manifests as gastrointestinal polyposis and mucocutaneous pigmentation, a more significant risk for various cancer types. The diagnostic criteria for PJS include the presence of two or more histologically confirmed PJ polyps, any number of PJ polyps in individuals with a positive family history of PJS, or in an individual who also has characteristic mucocutaneous pigmentation. Hamartomatous polyps, commonly manifest when a person is still relatively young, are also a part of PJS. Polyps typically first emerge when a person is 11-years-old. However, PJS was ruled out during the diagnosis because there was no unfavourable family history. No colonoscopy examination was done in this patient.

AD commonly presents with mucocutaneous discolouration. The skin, mouth cavity, conjunctiva, and genitalia can all be impacted by pigmented lesions. Brown spots on the tongue, palate, buccal mucosa, gingiva and gingiva are still other early signs of AD; however, unlike LHS, these spots are more dispersed and involvement of the palmar creases and flexural sites on the face where they are more evident because they are subject to pressure and light. Systemic manifestations of AD include anorexia, lack of energy, nausea, vomiting, weight loss, stomach and muscle discomfort, and orthostatic hypotension. In addition, AD patients may also have anaemia, lymphocytosis, eosinophilia, hypercalcemia, hyperkalaemia, hyponatremia, hypoglycaemia and high ACTH levels. For this patient, except for the pigmentation of the oral cavity, she did not have any other related symptoms. The blood investigations were also normal, ruling out AD.

The other differential diagnosis of mucocutaneous pigmentation should consider rare genetic syndromes like McCune-Albright syndrome, LEOPARD (lentigines, electrocardiographic conduction abnormalities, ocular hypertelorism, pulmonary stenosis, abnormalities of the genitalia, retardation of growth, deafness), and LAMB (lentigines, atrial myxomas, mucocutaneous pigmentation). Again, except for hyperpigmentation, the patient has no symptoms of the syndromes above.

Up to now, no evidence supports that there is a malignant tendency associated with LHS. It seems that there is no

systemic abnormality or familial factor associated with the syndrome. Generally, pigmentary changes in individuals with LHS do not disappear naturally but slowly increase along with aging. There are, therefore, no justifications for LHS therapy other than aesthetic and psychological components. Successful removal of pigmented lesions using laser techniques was reported[8] and to prevent the lesions from reoccurring, it is advisable to avoid the sun.[9]

In adolescents especially female, psychosocial assessment is warranted during follow-up to identify stress and anxiety in relation to unreasonable worry about the cosmetic appearance of the oral lesion. Reassurance and parental empowerment with correct knowledge is the key.

CONCLUSION

When making a differential diagnosis for mucocutaneous pigmentation, primary care doctors should always consider Laugier-Hunziker syndrome (LHS), particularly if the patient has no systemic symptoms. Despite its low prevalence, a rapid clinical diagnosis will allow the exclusion of more severe pigment illnesses and avoid the need for an additional test, intrusive examinations, and treatments. Specific investigation to establish the diagnosis is still unknown, but a more significant challenge in primary care is in identifying the lesion and establishing the cause of hyperpigmentation. It's important to underline the importance of promptly detecting LHS in young patients as early detection and diagnosis in this group of patients may prevent the emergence of mental health issues such as stress and anxiety connected to the uncertainty of the condition. Although most pigmentation disorders are benign or nonspecific, some disorders of skin pigmentation present cosmetic or psychological challenges to the patient, necessitating evaluation and treatment. Proper diagnosis of these common skin conditions will allow the physician to facilitate appropriate skin treatment and reassure the patient.

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CONFLICT OF INTEREST

None to declare.

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