CASE REPORT

Laugier-Hunziker syndrome: A case report

Manish Rijhwani, MD, Aisha A. Kh. M. Alkhadhari, MD

Department of Dermatology, Farwaniya Hospital, Ministry of Health, Kuwait

ABSTRACT

We describe a 47-year-old woman referred to Laser department for the management of hyperpigmented macules over oral musosa. The patient reported having these changes for years with no symptoms. A diagnosis of the Laugier-Hunziker syndrome (LHS) was made.

The Laugier-Hunziker syndrome (LHS) is an acquired, benign pigmentary skin condition involving the oral mucosa, often associated with longitudinal melanonychia. It is a diagnosis of exclusion, and other systemic conditions should be excluded prior to making a diagnosis. From the original description to date, around 200 cases have been reported world wide, mostly in Whites and published in the dermatological field. To our knowledge, the current report is the first report from Kuwait.

CASE REPORT

A 47-year-old woman presented with multiple pigmented macules dispersed over the oral and lip mucosa (Fig. 1, 2). The patient denied any symptoms related to any of these lesions. On examination, irregular brown spots with smooth and flat surface were observed on the left index finger and right index and ring fingers (Fig. 3). No fingernail lesions were observed. Our patient neither had a history of chronic drug use nor had familial history of pigmentary disorders and digestive polyposis or tumors.

Laboratory investigation results, including a full blood count, hematinic levels, serum chemistry and inflammatory markers, were all within normal range. Her hepatic and renal functions were normal and serum triiodothyromine (T3), tetraiodothyronine (T4) and thyrotropin (TSH) were all within normal limits. To rule out Addison's disease, the serum, urine cortisol, and

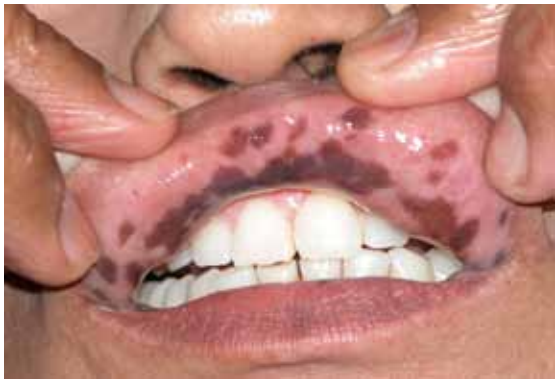

Fig. 1 Showing pigmented macules on the upper lip mucosa.

Fig. 2 Showing pigmented macules on the lower lip mucosa.

Correspondence: *Dr. Manish Rijhwani, Department of Dermatology, Farwaniya Hospital, Ministry of Health, Kuwait*

adrenocorticotrophic hormone (ACTH) levels were requested and all were within normal levels. Other possible causes of pigmentation such as drugs or metals were excluded by detailed history. Upper gastrointestinal endoscopy and colonoscopy were considered but not done as the patient did not have any sign or symptom of intestinal disease, and the patient's age and the absence of family history were deemed in contrast with the possibility of Peutz-Jeghers syndrome. The serology tests for human immunodeficiency virus (HIV) gave negative results. Because she was in good health, no further examinations were conducted and the patient refused for biopsy from the oral mucosa. A diagnosis of LH syndrome was made based on the clinical presentation of lesions coupled with the absence of systemic findings.

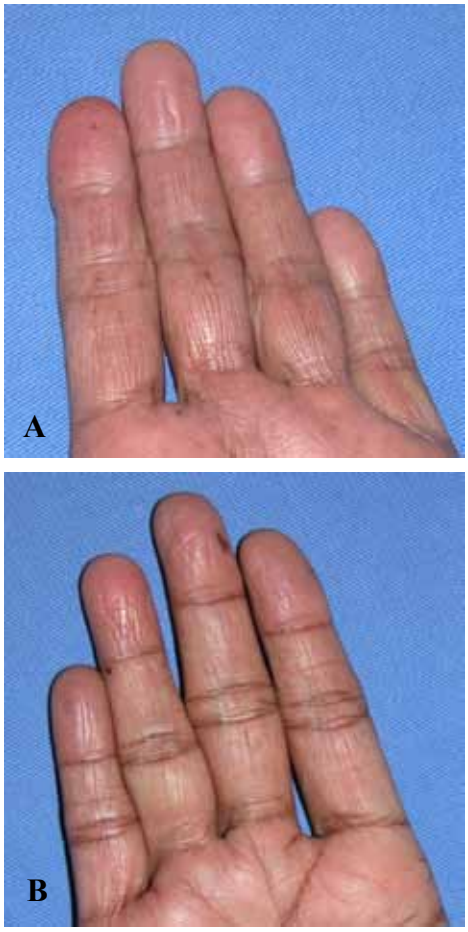

Fig. 3 A & B: Showing multiple small irregular brown macules on palmar aspect of fingers.

No systemic symptoms are associated with this syndrome. Prompt clinical recognition also averts the need for excessive and invasive procedures and treatments. Normally, no treatment is required for this condition, unless for aesthetic reason, mainly due to pigmentation on the lip mucosa.

DISCUSSION

Laugier–Hunziker syndrome (LHS), also known as idiopathic lenticular mucocutaneous pigmentation, was first described in 1970.[1] It is an acquired pigmentary condition affecting lips, oral mucosa and acral area, frequently associated with longitudinal melanonychia. There is neither malignant predisposition nor underlying systemic abnormality associated with LHS.[2]

LHS is characterized by a varying number of asymptomatic, lenticular (lens-shaped), or linear, brown to black mucocutaneous macules, usually less than 5 mm in diameter. They may be single or confluent. They may have well-defined or have indistinct margins. The hyperpigmentation occurs spontaneously and gradually and it is considered permanent.[1,3] The lesions are most commonly located on the lips, buccal mucosa, and hard palate. The less frequently affected areas include the soft palate, tongue, gingiva and floor of the mouth. Not all patients have both oral and nail involvement, the incidence of a pigmented nail band in cases of LHS is 44%–60%.[4,5]

LHS occurs predominantly among middle-aged adults with a mean onset at 50 years of age and occurrence is usually seen after puberty. It is more prevalent in women and most reported cases have been in Whites, particularly in French and Italians.[1,6] The diagnosis of LHS is frequently made clinically and is a diagnosis of exclusion.[7] Biopsy of the lesions can be performed to confirm

the diagnosis, and histopathologic changes associated with LHS show increased basal layer pigmentation with a normal number and morphologic appearance of melanocytes.[8]

The pathogenesis is thought to be linked to a functional alteration of the melanocytes that induces increased synthesis of melanosomes and subsequent transport to the basal cell layers. The etiology of the same is unknown.[3]

Prior to making the diagnosis of LHS, it is important to exclude other associated systemic conditions including the Peutz-Jeghers syndrome (PJS) and Addison's disease. PJS is an autosomal dominant genetic disorder that shares clinical features with LHS and is an important differential diagnosis to exclude due to its increased risk of malignancy.[9] PJS is characterized by hamartomatous gastrointestinal polyposis and hyperpigmentation of the skin and mucous membranes and is associated with an increased risk of both intestinal and extraintestinal malignancies. Diagnosis of PJS is made based on the presence of polyps in the gastrointestinal tract and a family history of the disorder, neither of which was present in our patient. The pigmented macules in PJS usually occur in infancy or early childhood, reaching a maximum at puberty.[9] The lesions of LHS, however, are progressively acquired after puberty. The pigmented lesions of the LHS are usually confined to the oral mucosa, lips, and nails, whereas PJS is often seen on the hands and feet.

Addison's disease, an endocrine disorder caused by insufficient production of cortisol and aldosterone, is characterized by increased pigmentation of the knuckles, skin creases, and mucous membranes[10] and is associated with increased level of circulating adrenocorticotropic hormone (ACTH). Other systemic findings include hypotension, dehydration, and abdominal pain, which were all absent in our patient.

Other differential diagnosis included McCune–Albright syndrome, drug-induced pigmentation, effects of smoking, heavy metal exposure, lichen planus and acquired immune deficiency syndrome (AIDS).

The most common drugs associated with oral pigmentation are tetracyclines, especially minocycline, antimalarials (chloroquine, hydroxychloroquine), amiodarone, chemotherapeutics, clofazamine, oral contraceptives, phenothiazines, azidothymidine, zidovudine and ketoconazole. Drug-induced oral pigmentation usually occurs following long-term use over months and years and often resolves after discontinuation of the causative drug.[11]

Treatment for the hyperpigmented macules in LHS is sought mainly for cosmetic reasons and includes cryosurgery, Q-switched Nd:YAG and Q-switched alexandrite laser therapy. Sun protection is important to prevent reoccurrence.[8]

Negative evidence of systemic symptoms such as fatigue, weight loss, cardiovascular, or gastrointestinal disorders, normal plasma levels of cortisol and ACTH, negative drug history, and negative findings in upper gastrointestinal endoscopy and colonoscopy will aid in the diagnosis of LHS. Hence, detailed history taking and through clinical examination of a patient presenting with oral pigmentation is of paramount importance.[12]

LHS is probably more common than is generally recognized.[4] We should be more familiar with the disorder. Laugier-Hunziker syndrome should always be considered in the differential diagnosis of a middle-aged patient presenting with mucocutaneous and ungual hyperpigmentation but

no systemic signs or symptoms. By recognizing LHS in the clinical setting, other pigmentary diseases can easily be excluded, and unwarranted tests and procedures can be prevented.

REFERENCES

1. Pereira PM, Rodrigues CA, Lima LL, Reyes SA, Mariano AV. Do you know this syndrome? An Bras Dermatol. 2010; 85 (5):751-53.
2. Int J Oral Sci. 2012 Dec; 4(4): 226–230. Laugier–Hunziker syndrome: a report of three cases and literature review
3. Siponen M, Salo T. Idiopathic lenticular mucocutaneous pigmentation (Laugier-Hunziker syndrome): A report of a case. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2003; 96:288-92.
4. Yago K, Tanaka Y, Asanami S. Laugier–Hunziker–Baran syndrome. Oral Surg Oral Med Oral Pathol Oral RadiolEndod. 2008; 106 2:e20-e25.
5. Baran R. Longitudinal melanotic streaks as a clue to Laugier–Hunziker syndrome. Arch Dermatol. 1979;115 12:1448-49.
6. Jabbari A, Gonzalez ME, Franks AG, Jr, Sanchez M. Laugier Hunziker syndrome. Dermatol Online J. 2010; 16:23.
7. Nayak RS, Kotrashetti VS, Hosmani JV. Laugier-Hunziker syndrome. J Oral Maxillofac Pathol. 2012; 16 (2):245-50.
8. Sachdeva S, Sachdeva S, Kapoor P. Laugier-Hunziker syndrome: a rare cause of oral and acral pigmentation. J CutanAesthet Surg. 2011; 4 (1):58-60.
9. Lampe AK, Hampton PJ, Woodford-Richens K, Tomlinson I, Lawrence CM, Douglas FS. Laugier-Hunziker syndrome: an important differential diagnosis for Peutz-Jeghers syndrome. J Med Genet. 2003; 40 (6):e77.
10. Wang WM, Wang X, Duan N, Jiang HL, Huang XF. Laugier-Hunziker syndrome: a report of three cases and literature review. Int J Oral Sci. 2012; 4 (4):226-30.
11. Zuo YG, Ma DL, Jin HZ, et al. Treatment of Laugier–Hunziker syndrome with the Q-switched alexandrite laser in 22 Chinese patients. Arch Dermatol Res. 2010;302 2:125-30.
12. Ko JH, Shih YC, Chiu CS, Chuang YH. Dermoscopic features in Laugier-Hunziker syndrome. J Dermatol. 2011; 38:87-90.