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CASE REPORT

Laugier-Hunziker syndrome: A diagnostic dilemma?

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Abstract: Laugier-Hunziker syndrome (LHS) is an idiopathic hypermelanotic condition that displays a characteristic pattern of mucosal, acral and nail pigmentation. The etiology is unknown, while its benign nature has been repeatedly highlighted. Owing to close resemblance to more serious disorders, it is necessary that the diagnostic features are understood; and thereby we report two sporadic cases of LHS in different age groups displaying varied presentations with identifiable features.

Keywords: Laugier-Hunziker syndrome; Hutchinson's sign

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Introduction

Laugier-Hunziker syndrome (LHS) is an idiopathic hypermelanotic condition that displays characteristic mucosal, acral and nail pigmentation[1]. The etiology is unknown, while its benign nature has been repeatedly highlighted[1-3]. Previous literatures have outlined a total of 180 cases merely as case reports; however, being a diagnosis of exclusion, unrecognized cases seem likely[4]. However, owing to its close resemblance to more serious disorders, it is necessary that a correct diagnosis be made. Herewith we report sporadic cases of LHS in different age groups with varied presentation.

Case reports

Case 1

A 14-year-old girl presented with asymptomatic lip pigmentation since 6 years of age. She also noted darkening of nails since last 4 months. There was no history of drug intake, similar family history or systemic features. Examination revealed multiple hyperpigmented macules over lips, buccal mucosa, tongue and dorsa of hands with pigmentary bands and homogenous pigmentation of fingernails were discerned as seen in **Figures 1 A–F**. Blood pressure, blood chemistry, serology (HIV) and thyroid function tests were within normal limits. Ultrasonography showed normal findings. The patient was counseled and reassured of the benign course.

Case 2

A 55-year-old man presented with asymptomatic dark macules over the feet since 2 years, hands for 6 months, and nail discoloration since 2 years. Initial small pinpoint lesions progressed up to 1 cm in size. There was no history of tobacco addiction, drug intake, similar lesions in family members or systemic complaints. On examination, multiple well-defined light-brown-to-black macules up to 1 cm in size were observed over labial mucosa, buccal mucosa, hard palate, distal phalanges of hands, soles, and prepuce of penis, with varied patterns of nail pigmentation, as noted in **Figures 2 A–E**. A few macules involved the proximal and lateral nail folds exhibiting a pseudo-Hutchinson's sign. An evaluation of vitals, blood chemistry, serology (HIV), ultrasound and upper gastrointestinal endoscopy revealed normal findings. Despite relief over its benign nature, the extensive evaluation and lack of affordable therapeutic options were not comforting to the patient.

Discussion

A classical description by Laugier and Hunziker in the year

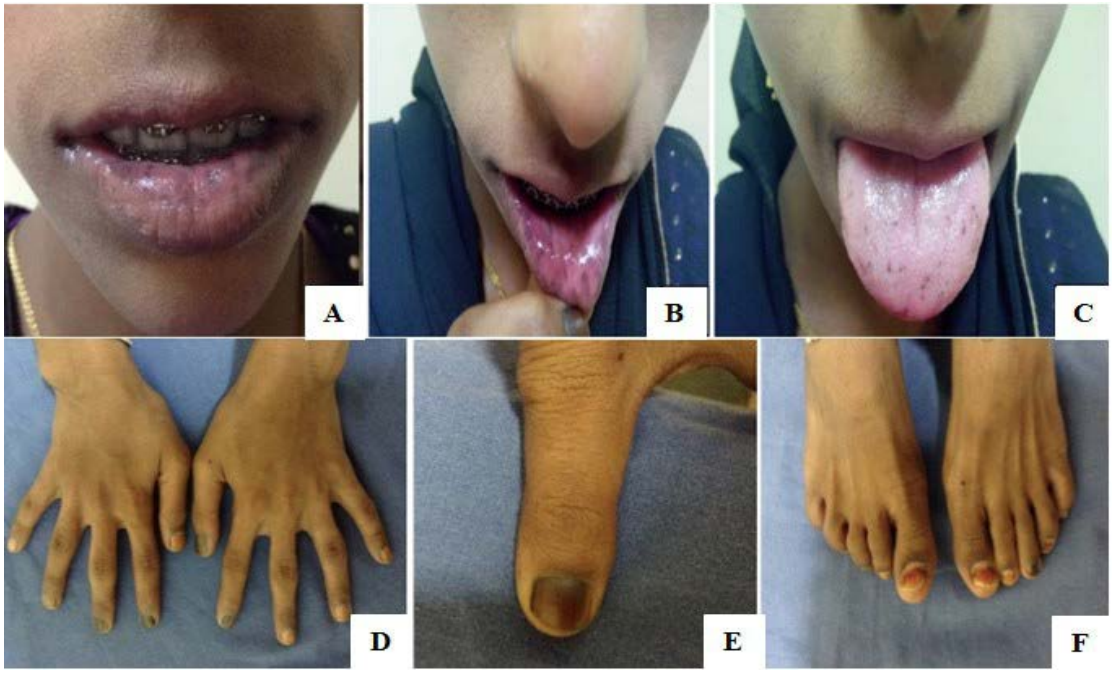

Figure 1. (A)–(C) Multiple discrete dark brown macules over lips, labial mucosa, and dorsum of tongue. (D)–(F) Varied pigmentation of nails.

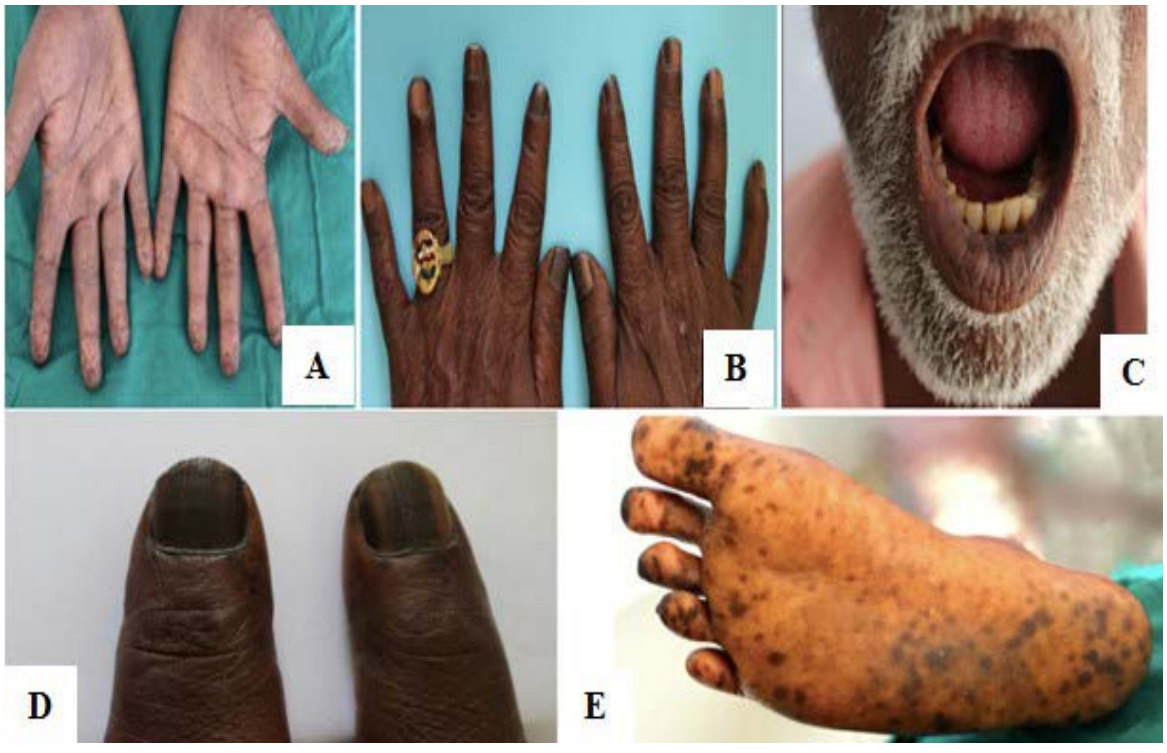

Figure 2. (A) Well-defined dark brown to black macules grouped over distal phalanges of hands. (B) Varied patterns of nail pigmentation with a few discrete hyperpigmented macules on dorsal hands. (C) A few dark brown macules on lower lips. (D) Discoloration and longitudinal ridging of nails with pseudo-Hutchinson's sign. (E) Brownish black macules and patches over the sole.

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1970 under the term "idiopathic lenticular mucocutaneous syndrome" was light-to-dark-brown blotchy macules less than 5 mm, limited to fingertips, soles and mucosal surfaces, with striking involvement of nail folds exhibiting a pseudo-Hutchinson's sign[5,6]. The nail involvement is described as four types: single 1–2 mm longitudinal streaks, double 2–3 mm lateral longitudinal streaks, radial or ulnar half homogenous pigmentation, and complete pigmentation[7]. A similar presentation is seen herewith, with more prominent involvement in older age. It may be known that varied presentations have been reported previously as isolated tongue, neck and trunk pigmentation, however, these are rare[4,7–11].

A higher incidence in Chinese population follows a genetic influence of either autosomal dominant or, less commonly, autosomal recessive; in contrast, isolated sporadic case reports occur in other Asian continents[6,12]. The proposed mechanism of LHS is the presence of altered melanocytes, thereby leading to increased melanogenesis[13]. Findings of basal layer pigmentation, large dendritic L-3, L-4 dihydroxyphenylalanine reactive intraepithelial melanocytes, and a few dermal melanophages with electron microscopic features of multiple mature melanosomes within keratinocytes and melanophages, confirm the benign nature[14]. Despite its benign nature, there are rare associations such as esophageal melanocytosis, actinic lichen planus, hypocellular bone marrow and thrombocytopenia that may be kept in mind and does not necessitate evaluation[8].

The disorders closely simulating LHS by appearance are Addison's disease, Peutz-Jeghers syndrome, and lentiginosis profuse[8]. The features differentiating Laugier-Hunziker syndrome from other disorders are striking by mere clinical evaluation, as outlined in **Table 1**. Nonetheless, majority of cases are reported following repeated invasive procedures such as colonoscopy, gastroscopy and barium enema[11].

As for both cases outlined herewith, no treatment was necessary due to absence of cosmetic concern and summed up to reassurance alone. This outlook has been favorable according as Ergun *et al.* highlighted the resistant nature with recurrence following laser therapy[15].

Conclusion

Despite the strikingly unique features of LHS, the categorization of it as a diagnosis of exclusion may be noted as leading to a battery of investigations, thereby causing undue anxiety upon patients. It may thus seem necessary to make aware that the disorder is relatively

Table 1. Differentiating features of disorders exhibiting mucosal and cutaneous hyperpigmentation[7,8,11,12].

	Laugier-Hunziker syndrome	Peutz-Jeghers syndrome	Addison's disease	Cronkhite-Canada syndrome	Lentiginosis profusa
Mechanism	Hyperactive melanocytes	Germline mutation in *STK11/LKB1* tumor suppressor gene on chr 19p13.3	Excess adrenocorticotrophic hormone stimulates melanocytes.	Protein losing enteropathy	Mutations in gene for protein-tyrosine phosphatase, non-receptor type 11
Inheritance	Sporadic/Autosomal dominant	Autosomal dominant	Nil	Nil	Autosomal dominant
Onset of lesions	Early to middle adulthood	Childhood	Variable, insidious	Adult	At birth
Skin manifestations	Multiple, 1–5 mm light-to-dark-brown macules over distal aspects of digits and soles	1–5 mm dark brown macules at perioral, nose, digits, hands, feet and perianal sites	Generalized hyperpigmentation; more pronounced on sun-exposed areas, palmar creases, knuckles, elbows, knees and scars	Multiple light-to-dark-brown macules on extremities, face, palms, soles	Multiple brown macules and patches up to several cm in size, involving around 80% of body surface area
Mucosal features	1–5 mm hyperpigmented macules involving labial, buccal, gingival, palatal, tongue and genital mucosae	1–5 mm hyperpigmented macules involving buccal and gingival mucosae	Hyperpigmented patches over dentogingival margins, buccal, vaginal and perianal mucosae	Usually spared. Buccal mucosa occasionally affected.	Multiple brown macules on buccal mucosa and sclera
Nail changes	Longitudinal melanonychia or homogenous pigmentation	Clubbing	Nail bed pigmentation	Dystrophic changes	Occasional longitudinal pigmentary bands
Associations	Esophageal melanocytosis, actinic lichen planus, hypocellular bone marrow and thrombocytopenia.	Intestinal polyposis, malignancy	Nausea, vomiting, diarrhea, steatorrhea, dizziness, myalgia and arthralgia	Gastrointestinal polyposis and alopecia	ECG changes, ocular hypertelorism, pulmonary stenosis, abnormal genitalia, retarded growth and deafness

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common among both juveniles and adults with no plausible hereditary component.

Conflict of interest

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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