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 1A–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 2A–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.
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. 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. 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.

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. The proposed mechanism of LHS is the presence of altered melanocytes, thereby leading to increased melanogenesis. Findings of basal layer pigmentation, large dendritic melanocytes, thereby leading to increased melanogenesis, confirm the microscopic features of multiple mature melanosomes within keratinocytes and melanophages, causing undue anxiety upon patients. It may thus seem necessary to make aware that the disorder is relatively benign nature. 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.

The disorders closely simulating LHS by appearance are Addison’s disease, Peutz-Jeghers syndrome, and lentiginosis profusa. 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.

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.

### 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 benign nature. 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.

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.

### Table 1. Differentiating features of disorders exhibiting mucosal and cutaneous hyperpigmentation.

<table>
<thead>
<tr>
<th></th>
<th>Laugier-Hunziker syndrome</th>
<th>Peutz-Jeghers syndrome</th>
<th>Addison’s disease</th>
<th>Cronkhite-Canada syndrome</th>
<th>Lentiginosis profusa</th>
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<tbody>
<tr>
<td><strong>Mechanism</strong></td>
<td>Hyperactive melanocytes</td>
<td>Germline mutation in STRK1/LKB1 tumor suppressor gene on chr 19p13.3</td>
<td>Excess adrenocorticotropic hormone stimulates melanocytes.</td>
<td>Protein losing enteropathy</td>
<td>Mutations in gene for protein-tyrosine phosphatase, non-receptor type 11</td>
</tr>
<tr>
<td><strong>Inheritance</strong></td>
<td>Sporadic/Autosomal dominant</td>
<td>Autosomal dominant</td>
<td>Nil</td>
<td>Nil</td>
<td>Autosomal dominant</td>
</tr>
<tr>
<td><strong>Onset of lesions</strong></td>
<td>Early to middle adulthood</td>
<td>Childhood</td>
<td>Variable, insidious</td>
<td>Adult</td>
<td>At birth</td>
</tr>
<tr>
<td><strong>Skin manifestations</strong></td>
<td>Multiple, 1–5 mm light-to-dark-brown macules over distal aspects of digits and soles</td>
<td>1–5 mm dark brown macules at perioral, nose, digits, hands, feet and perianal sites</td>
<td>Generalized hyperpigmentation; more pronounced on sun-exposed areas, palmar creases, knuckles, elbows, knees and scars</td>
<td>Multiple light-to-dark-brown macules on extremities, face, palms, soles</td>
<td>Multiple brown macules and patches up to several cm in size, involving around 80% of body surface area</td>
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<tr>
<td><strong>Mucosal features</strong></td>
<td>1–5 mm hyperpigmented macules involving labial, buccal, gingival, palatal, tongue and genital mucosa</td>
<td>1–5 mm hyperpigmented macules involving buccal and gingival mucosa</td>
<td>Hyperpigmented patches over dentogingival margins, buccal, vaginal and perianal mucosa</td>
<td>Usually spared. Buccal mucosa occasionally affected.</td>
<td>Multiple brown macules on buccal mucosa and sclera</td>
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<td><strong>Nail changes</strong></td>
<td>Longitudinal melanonychia or homogenous pigmentation</td>
<td>Clubbing</td>
<td>Nail bed pigmentation</td>
<td>Dystrophic changes</td>
<td>Occasional longitudinal pigmented bands</td>
</tr>
<tr>
<td><strong>Associations</strong></td>
<td>Esophageal melanocytosis, actinic lichen planus, hypocellular bone marrow and thrombocytopenia.</td>
<td>Intestinal polyposis, malignancy</td>
<td>Nausea, vomiting, diarrhea, steatorrhea, dizziness, myalgia and arthralgia</td>
<td>Gastrointestinal polyposis and alopecia</td>
<td>ECG changes, ocular hypertelorism, pulmonary stenosis, abnormal genitalia, retarded growth and deafness</td>
</tr>
</tbody>
</table>

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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.

**References**


