Journal of Chitwan Medical College 2020;10(32):87-89
Available online at: www.jcmc.cmc.edu.np

VIEW POINT

PROPOSED DIAGNOSTIC CRITERIA AND CLASSIFICATION FOR LAUGIER-HUNZIKER SYNDROME

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Received: 16 Mar, 2020
Accepted: 6 Jun, 2020
Published: 25 Jun, 2020

Key words: Classification; Diagnostic criteria; Laugier-Hunziker syndrome.

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*DOI:*https://doi.org/10.3126/jcmc.v10i2.29684

Citation

Mishra N. Proposed diagnostic criteria and classification for Laugier-Hunziker Syndrome.Journal of Chitwan Medical College.2020;10(32):87-89.

ABSTRACT

Laugier-Hunziker syndrome is one of those rare syndromes which has yet not been reviewed and reported much in literature. So far, there is no diagnostic criteria and classification that can help with prompt diagnosis of this syndrome. In this article an attempt has been made to put forward a simple criterion along with a classification that can assist the practitioners with early detection and also will have substantial contribution in medical curriculum.

INTRODUCTION

Laugier-Hunziker syndrome is named after the two scientists P. Laugier and N. Hunziker, who reported the first 5 cases of this syndrome in 1970. It was then termed as "Idiopathic lenticular mucocutaneous syndrome," as the cases clinically presented with acquired pigmentation of oral and genital mucosa with only 2 patients presenting additionally with nail pigmentation.[1] Later, in 1979 Robert Baran also reported 9 similar cases out of which 4 showed longitudinal melanotic streaks in nails. The mucocutaneous pigmentation in this syndrome is usually seen as small grey to dark brown colored macules ranging in size from 0.5-5mm.[2] Along with pigmentation of mucosa and skin; one or more finger or toe nail is frequently seen to be involved with exhibition of a wide variety of patterns of pigmentation but without dystrophy. Melanonychia in Laugier-Hunziker syndrome as described by Baran, can be seen in 3 different forms: single 1-2 mm wide longitudinal streaks; 2-3 mm wide double longitudinal streaks involving lateral nail plate; radial or ulnar half homogenous pigmentation,[1] and a 4th variant i.e. complete pigmentation of nail was later added by Veraldi et al in 1991.[2]

Even though the cases of Laugier-Hunziker syndrome have seen to be reported in the medical literature with an increasing frequency within the past few years; there are no strict criteria to diagnose and categorize it. The lack of criteria usually results in indecision or misdiagnosis along with non-essential clinical trials on patient.

After a thorough review of the literature[3-11]; I have made an attempt to highlight the criteria for prompt diagnosis of this syndrome along with drafting of a simple classification for the same.

DISCUSSION

"*Small, Simple yet Significant*"; that is how I define Laugier-Hunziker syndrome. A simple melanotic pigmented disorder, with small macules ranging from few millimeters to few centimeters, still it holds so much value when it comes to differentiating it from other similar looking but more worrying pigmented disorders.

Every year new cases of Laugier-Hunziker syndrome are reported and added to the medical literature, which so far have shown a wide and varied range of clinical presentation. But, non-existence of any accurate universal diagnostic criteria is

accountable for the lack of conviction resulting in a lot of controversies and doubt for a confirmatory diagnosis to be made in cases of this syndrome. I hereby stress on the need for a diagnostic criterion to be made and universally accepted for its firm diagnosis and also spreading awareness amongst the dentists as well as dermatologists in particular so that no case goes unnoticed.

So considering the exigency, after an intense study of the literature[1-11] I am hereby proposing a clinicopathologic diagnostic criteria (Table 1) and classification (Table 2), for Laugier-Hunziker syndrome that can assist the clinicians with its instant identification and categorization; and which can further be beneficial in researches and academics.

Table 1: Diagnostic criteria based on clinical and histological features

Clinical Criteria:	
Skin/Mucosa	a)Irregular lenticular hyperpigmented macules
	b)2-5 mm diameter
	c)Slate to Dark brown in color
	d)Well defined or indistinct margins
	e)Macules can be single or in multiple groups, sometimes confluent to form homogenous patches
	f)Commonly involved intraoral sites: lower lip, upper lip, tongue, buccal mucosa, hard and soft palate
	g)Commonly involved extraoral sites: palmoplantar area, fingertips, genital region
Nail Plate* (As given by Baran)	a)Single 1-2mm, longitudinal streaks
	b)Double 2-3mm, longitudinal streaks
	c)Radial or Ulnar half homogenous pigmentation
	d)Complete pigmentation
Histological Criteria:	
a)Increased melanin pigment in basal cell layer	
b)Normal number and morphology of melanocytes	
c)Increased number of melanophages in submucosa or papillary dermis	
d)Dermal pigmentary incontinence	

*One or more fingernails or toenails can be affected with discrete longitudinal hyperpigmented bands of varying width and intensity but without any associated nail dystrophy.

Table 2:Clinical classification depending on the site of involvement

Class 1: Lip, Oral Mucosa and Nail	
Type I	Involvement of Lip and Oral mucosa without Nail involvement
Type II	Involvement of Lip and Nail* without involvement of Oral mucosa
Type III	Involvement of Oral mucosa and Nail* without involvement of Lip
Type IV	Involvement of Lip, Oral mucosa, Nail, Perioral region, Palmoplantar area and Skin
Class 2: Mucocutaneous Pigmentation involving Lip, Nail, Oral Mucosa, Skin with any other associated disorder	

* One nail, multiple nails or Involvement of Nail fold (Pseudo-Hutchinson's sign)

CONCLUSION

As per the basic concern, it is always important to first correctly identify the condition. And the present proposed illustration has the capability to reduce both the over and under diagnosis of this syndrome, thereby eliminating the uncertainty that clinicians face while categorizing the patients into different conditions.

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