

Nevus Unius Lateris with Bilateral Oral Mucosal Lesions: An Unusual Presentation

Abstract

Verrucous epidermal nevi (VEN) are cutaneous hamartomas characterized by keratinocytic hyperplasia. Majority are linear in distribution and tend to follow the Blaschko lines; however, some may have zosteriform (segmental) or systematized distribution involving widespread areas of skin. The systematized ones are further classified into “*Nevus Unius Lateris*” when one-half of the body is affected, and “*Ichthyosis Hystrix*” showing bilateral distribution, both being the uncommon forms. Although it can affect any body part, it rarely involves the head and neck region with seldom involvement of mucosae, scalp, and ear lobes. We saw a 6-year-old child with multiple hyperpigmented verrucous plaques predominantly present over left half of the body, ipsilateral alopecia scalp, and verrucous lesions involving mucosae of palate and tongue, which were present bilaterally. Previously, case reports of oral lesions related to VEN had demonstrated segmental, midline, or unilateral distribution. Hereby, we report this peculiar case of *Nevus Unius Lateris* with bilateral oral mucosal involvement, owing to its rarity.

Keywords: *Cutaneous mosaicism, nevus unius lateris, oral epidermal nevus, verrucous epidermal nevus*

Introduction

Verrucous epidermal nevi (VEN) are cutaneous hamartomas which originate from the embryonic ectoderm. The onset is usually at birth or early life with a prevalence of 1:1000 live births.^[1] They are characterized by skin-colored to brown-gray papules which may coalesce to form plaques arranged in linear-streaks and swirls which do not generally cross the midline.^[2] Presentation may be localized or widespread with different patterns such as linear, zosteriform and rarely, diffuse or systematized. When it involves extensive body surface area, the term “Systematized Epidermal Nevus” is preferred. Its variants include “*Nevus Unius Lateris*” when it covers one-half of the body and “*Ichthyosis Hystrix*” when distributed bilaterally.^[3]

Case Report

A 6-year-old female presented to Dermatology Outpatient Department with cosmetically disfiguring asymptomatic lesions in form of hyperpigmented verrucous plaques predominantly covering

the left half of the body extending from scalp, face, external ear, neck, trunk, labium majus, left upper and lower limbs till the dorsum of left foot [Figure 1a-c]. Few thin streaks were also present over contralateral side of mid-face (lower eyelids, cheeks, upper lips), neck, shoulder [Figure 2a and b], and upper limb. The onset of the lesions occurred during infancy, and since then the lesions became darker, raised, and thickened with loss of hair over ipsilateral scalp. No history suggestive of developmental/milestones delay, seizures, mental retardation, and skeletal abnormality was present. Similar history in family was not present.

On examination, the hairs over the left half of scalp were thin, hypopigmented, shorter in length, and sparse in distribution characteristically along the distribution of nevus with rest of the scalp having well-developed terminal hairs [Figure 3a-c]. Lesions over flexural surfaces such as right axilla, neck, cubital fossa and knuckles were more corrugated than other sites. On palpation, the lesions were soft and velvety to touch. Knuckles and volar

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aspect of left hand and dorsum of left great toe had multiple, thick, hyperkeratotic plaques [Figure 4a-c]. Oral examination revealed gray-colored tiny papules arranged in linear clusters over bilateral superior vermillion, left oral commissure, and adjoining buccal mucosa. Intraorally, papilliform mucosal plaques with irregular surface and borders involved midline and bilateral palatal mucosa, and dorsum of tongue [Figure 5a-d].

No systemic abnormality was detected. Routine investigations including electrocardiogram, ophthalmological examination, and Ultrasound abdomen were normal. Consent for CT-scan Brain was not given. Histopathology from upper back revealed massive hyperkeratosis, acanthosis, papillomatosis, and elongation of rete ridges. Increase in basal melanin was also evident [Figure 6a and b]. Hence, the diagnosis of *Nevus Unius Lateris* with bilateral oral mucosal-VEN and ipsilateral alopecia of scalp was made. Though alopecia was remarkable along the distribution of nevus, no signs of scarring were evident.

Discussion

VEN represent a form of cutaneous mosaicism which emerges as a result of de-novo postzygotic mutations.^[4] The common sites include trunk and extremities where lesions attain transverse and longitudinal configurations, respectively, following the Blaschko lines. It is uncommon over the head and neck region; rarely involves face, scalp, ear lobes^[2] and very rarely affects the

oral mucosa.^[5] In our case, apart from the involvement of these sites, oral lesions were also recorded.



Figure 1: "Nevus Unius Lateris" on (a) anterior trunk with sharp midline demarcation and predominant left upper-limb involvement; (b) Posterior trunk. Note lesions over nape of neck and left-axilla are more verrucous; (c) Left lower limb involvement

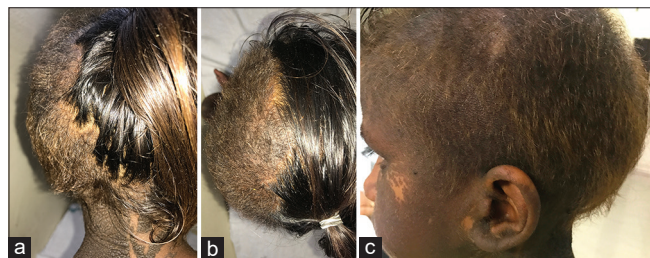


Figure 3: (a) Sparsely distributed hairs on left half of scalp with terminal hairs on contralateral side (posterior view); (b) Top view; (c) Lateral view with external ear involvement evident



Figure 2: (a) Predominant left side involvement of face with few streaks involving contralateral skin of lower-eyelid, cheek, and upper-lip; (b) Left upper-eyelid involvement; Note paucity of hairs on left eyebrow



Figure 4: Thick hyperkeratotic lesions involving (a) thenar eminence, palmar surfaces of left thumb, and base of index finger; (b) PIP joints of left 4th and 5th fingers; (c) Dorsum of base of left great toe

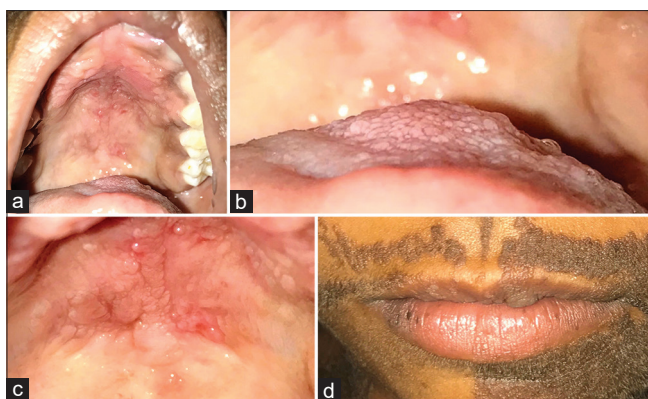


Figure 5: (a) Verrucous mucosal plaques on palate and dorsum of tongue; (b) Tongue lesions under magnification giving a cobblestone appearance; (c) Palatal mucosa under magnification; (d) Lip lesions. Note moustache-like pattern on upper lips with the extension of lesions from philtrum and philtral ridges to lip tubercle and both sides of superior vermilion respectively

Historically, Brown and Gorlin in 1960 gave the first review on oral manifestations in verrucous epidermal nevus. Since then, only 16 cases of oral-VEN with or without cutaneous epidermal nevus have been described in the worldwide literature over a period of 60 years [Table 1]. Out of these, only a single case reported by Bygum *et al.* had systematized distribution of nevus (*Ichthyosis Hystrix*),^[4] while nine cases had localized cutaneous involvement and six patients had isolated oral-VEN without any cutaneous feature.^[6,9-11] None of these patients had nervous, ocular or skeletal abnormalities, hence, did not fulfil the criteria of Epidermal Nevus Syndrome (ENS). On the other hand, there are multiple case reports in literature describing the oral manifestations in ENS. Worthy of note is the case of ENS described by Kelley *et al.* (1972) of a young boy having epidermal nevus involving the head and neck region. The boy also had blindness, mental retardation, and alopecia of scalp along the distribution of nevus. The oral lesions were highly disfiguring and mental deficiency led to factitious oral ulceration in this case.^[12]

Oral-VEN can involve any oral structure including lip vermilions, oral commissures, labial and buccal mucosa, gingivolabial sulcus, palatopharyngeal folds, hard or soft palate but they usually remain homo-lateral to their cutaneous counterparts. Their color may vary from normal-mucosa-like to yellow-white, tan, dark-brown, or gray. They usually present as unilateral/midline papules or sessile nodules with a verrucous, papillary, mammilated or condylomatous surface which characteristically do not cross the midline.^[5,6] However, the case reported by Bygum *et al.* of bilaterally systematized epidermal nevus had oral lesions which were also bilateral.^[4] Although our case was predominantly *Nevus Unius Lateris*, the bilateral oral lesions could be explained by their bilateral cutaneous counterparts over mid-face (specifically cheeks and upper lips). These topographical considerations may imply that lines of Blaschko, originally illustrated in skin, may have

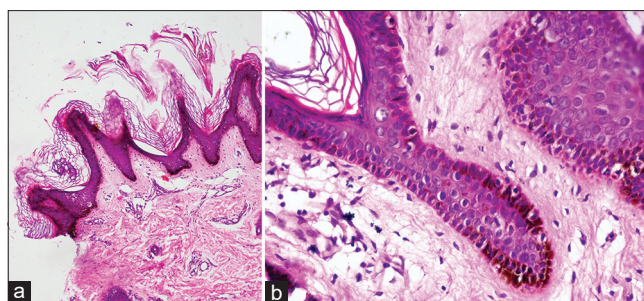


Figure 6: (a) Presence of extensive hyperkeratosis, irregular acanthosis, papillomatosis, and elongation of rete ridges (H and E, 100 \times); (b) High basal melanin content observed at higher magnification (H and E, 400 \times)

intraoral counterparts caused due to proliferation of oral clones of epithelial progenitor cells.^[9]

Histopathologically, oral-VEN shows papillomatous projections with a moderate degree of hyperkeratosis, acanthosis, and elongation of rete ridges which correlates clinically to the raised papillary lesions with a micropapillary or verrucous surface. The clinical diagnosis of oral-VEN is often supported by the presence of cutaneous lesions of VEN. However, the establishment of diagnosis becomes particularly difficult in the absence of cutaneous counterpart and differentiation from other oral diseases such as squamous papilloma, oral-verruca vulgaris becomes vital. Oral-VEN appears usually at birth or childhood, while squamous papilloma develops during 30–60 years of age. Also, squamous papilloma is usually small, pedunculated, and shows signs of keratinization with white surface, whereas oral-VEN is generally sessile, with the same color as normal mucosa.^[6] Oral verruca vulgaris in children may clinically resemble oral-VEN; to differentiate, the former are acquired; sessile lesions and enlarge rapidly in a short timeframe with stabilization to a maximum size of 4–5 mm, whereas the lesions in latter have the tendency to constantly enlarge until their phase of somatic growth is over. Histopathologically, verrucae show finger-like projections of hyperkeratotic stratified-squamous epithelium producing a “cupping effect” with or without koilocytes.^[9] In adults, it needs to be differentiated from verrucous carcinoma. The clinical correlation and history of onset and progression facilitate in ascertaining the correct diagnosis.

Treatment of oral-VEN lesions is generally not required if it does not cause any functional abnormality.^[7] Fortunately, oral lesions in our patient were not problematic; in fact, they remained unnoticed until our examination. Surgical excision of oral lesions has been tried but recurrence was noticed as they tend to regrow during their phase of somatic growth till adolescence.^[11] Considering possible malignant transformation which is well-documented in cutaneous VEN, no case of neoplastic transformation in oral-VEN is reported till date.^[7] Nevertheless, systematized-VEN are managed symptomatically with appropriate counselling for the significance of long-term follow-up as the risk of

Table 1: Data of clinical features of oral-VEN observed by different authors

Study (Year)	Case No.	Age	Sex	Presentation	Age of onset	Oral sites of involvement	Cutaneous sites of involvement
Present case (2020)	1	6 y	F	Left half of body, bilateral mid-face and oral mucosa	First year of life	Both sides of superior vermillion, left oral commissure, left side of buccal mucosa, hard palate, dorsum of tongue	Nevus unius lateris extending from scalp, neck, trunk, extremities, external genitalia with associated alopecia of scalp and left eyebrow
Maly et al. (2016) ^[7]	2	20 y	M	Right	Birth	Right side of upper lip vermillion, maxillary gingiva, hard palate	Linear lesions over right side of face
Santos et al. (2012) ^[8]	3	51 y	F	Left	Birth	Left hemipalate, superior labiogingival sulcus	Nasal columella, upper lip
Bygum et al. (2011) ^[4]	4	17 y	F	Bilateral	4 months of age	Bilateral lower labial mucosa, buccal mucosa close to oral commissures, hard palate laterally	Bilateral Systematized VEN (Ichthyosis Hystrix) whole body including scalp with no associated alopecia
Tesi and Ficarra (2010) ^[9]	5	32 y	M	Left	Child-hood	Left lower labial mucosa and border of tongue	Absent
Haberland	6	4 y	M	Midline	Child-hood	Midline hard palate	Absent
Carrodeguas et al. (2008) ^[6]	7	13 y	F	Midline, right	Birth	Midline hard and soft palate	Midline chin and right neck
	8	17 y	M	Right	Birth	Right buccal mucosa, maxillary vestibule, hard and soft palate, right side of dorsum of tongue	Right upper lip vermillion, and perioral skin
	9	6 y	M	Left	Birth	Left buccal mucosa, maxillary vestibule and attached gingiva	Left arm, chest, neck, perioral skin
	10	63 y	M	Right	Birth	Right anterior soft palate, midline hard palate	Right nasolabial area, upper lip vermillion
	11	4 y	F	Left	4 years (noticed first)	Left buccal mucosa, retromolar pad, soft palate and palate-pharyngeal fold	Absent
Ozcelik et al. (2005) ^[5]	12	13 y	F	Right	Birth	Right oral commissure, buccal mucosa, soft and hard palate, uvula enlargement	Right corner of mouth and cheek
Coley-Smith and Shaw (1996) ^[10]	13		F	Midline and left	Birth	Left buccal and palatal mucosa, uvula enlargement	Midline lower lip, left corner of mouth and cheek
	14		M	Midline	Birth	Midline dorsum of tongue, Right palatal mucosa and gingiva	Absent
Hickman et al. (1988) ^[11]	15	3 y	F	Left	3 years of age (noticed first)	Left upper gingiva and oral commissure	Skin below left eye
	16	17 y	M	Midline	Child-hood	Midline soft and hard palate	Absent
	17	15 y	F	Midline	Child-hood	Midline anterior hard palate	Absent

y=years

malignant transformation, though rare, cannot be ruled out otherwise.^[1]

Oral mucosal and scalp biopsy could not be performed in our case as parents denied the consent. This factor remains as a limitation. However, this case of *Nevus Unius Lateris* with alopecia scalp, genital and mucosal VEN in this young Indian girl is reported for its unusual combination of predominant unilateral cutaneous presentation and bilateral oral mucosal involvement.

Declaration of patient

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for their images and other clinical information to be reported in the journal. The parents understand that patient names and initials will not be published and due

efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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