

Systematized Verrucous Epidermal Nevus- A Case Report

B.C. Sharathkumar¹, N.S. Anitha²

¹Professor and HOD, Department of Dermatology, Venereology and Leprosy, Kempegowda Institute of Medical Sciences Hospital and Research Centre, Bengaluru, Karnataka. ²Resident, Department of Dermatology, Venereology and Leprosy, Kempegowda Institute of Medical Sciences Hospital and Research Centre, Bengaluru, Karnataka.

INTRODUCTION

Veracious epidermal nevus (VEN) is a keratinocyte hematoma, usually present at birth or can develop later in life.^{1,2} It commonly involves trunk and limbs. Less commonly, involved sites are head and neck; face is known to involve even more rarely.^{3,4,5} Clinical variants of VEN are zosteriform, linear, unilateral or systematized patterns with streaks and swirls.⁶ We report a case of girl who presented with extensive VEN causing a lot of disfigurement. Epidermal nevi are hamartomas of cutaneous structures arising from the embryonic ectoderm. The prevalence rate is 1 in 1000, without predilection to race and sex.^{7,8} They are further divided into nonorganoid (Keratinocytic) types and organoid types (due to hyperplasia of adnexal structures such as sebaceous glands, sweat glands and hair follicles).^{8,9} All epidermal nevi represents disorder of dermo-epidermal interactions and most cases are sporadic. Verrucous epidermal nevus is the most common type of keratinocyte nevus and derives its name from its keratotic and warty appearance. It may present at birth, develops later in infancy or very rarely appears in adolescence.²

PRESENTATION OF CASE

A 12-year-old girl presented to dermatology outpatient department with complaints of multiple raised dark coloured skin lesions over face, neck, scalp and left side of the back since birth. There was a history of darkening and thickening of the lesions with age. The child was born from second degree consanguineous marriage and family history was not significant. Her growth and development were normal for age and attained milestones appropriate for age. No relevant past history suggestive of any systemic illness.

On examination, discrete to confluent multiple hyperpigmented, hyperkeratotic thick verrucous papules and plaques were present diffusely over forehead, right upper eyelid, bilateral eyebrows, bilateral ears, left side of nose, bilateral alae nasi, bilateral cheeks, chin, submental area and anterior aspect of neck, with islands of normal skin. Few verrucous papules were interspersed among them. (Figure 1, 2, 3). Over scalp, thick hyperkeratotic plaque was present diffusely over right fronto temporoparietal region with thinning of hair and causing scarring alopecia in few places. (Figure 4) Largest verrucous plaque measuring about 10 x 8 cms seen over nape of the neck on left side (Figure 5). Hyperpigmented thick verrucous plaques were present over left side of the back (Figure 6).

Based on history and clinical examination, differential diagnosis of verrucous epidermal nevus and nevus sebaceous were made. Detailed systemic examination was done to rule out skeletal, ophthalmic and neurological involvement and it was found to be normal. Routine laboratory investigations were within the normal range. Computed tomography scan was done and there were no abnormalities. Biopsy was done and showed hyperkeratosis, papillomatosis and acanthosis with elongation of the rete ridges, features consistent with verrucous epidermal nevus.

Corresponding Author:

Dr. N. S. Anitha,

Resident,

*Department of Dermatology,
Venereology and Leprosy,
Kempegowda Institute of Medical
Sciences Hospital and Research Centre,
Bengaluru- 560004, Karnataka.*

E-mail: anithabhavana219@gmail.com

DOI: 10.18410/jebmh/2019/637

*Financial or Other Competing Interests:
None.*

How to Cite This Article:

*Sharathkumar BC, Anitha NS.
Systematized verrucous epidermal
nevus- a case report. J. Evid. Based Med.
Healthc. 2019; 6(48), 3048-3051. DOI:
10.18410/jebmh/2019/637*

Submission 20-11-2019,

Peer Review 25-11-2019,

Acceptance 30-11-2019,

Published 02-12-2019.



	<p>Figure 1. <i>Hyperpigmented, Verrucous Thick Plaques Over Face</i></p>
	<p>Figure 2. <i>Verrucous, Hyperpigmented Plaque in Submental Area and Neck</i></p>
	<p>Figure 3. <i>Thick Hyperpigmented, Verrucous Plaques Involving Left Side of the Face, Left Ear and Left Temporal Region</i></p>
	<p>Figure 4: <i>Hyperpigmented, Verrucous Plaques Involving Right Ear, Right Frontal, Parietal and Temporal Regions with Thinning of Hair. Scarring Alopecia Over Scalp and Lateral Aspect of Right Eyebrow.</i></p>
	<p>Figure 5. <i>Large Verrucous Plaque Over Nape of Neck</i></p>
	<p>Figure 6. <i>Hyperpigmented Plaques Over Back on Left Side</i></p>

CLINICAL DIAGNOSIS

Systematized verrucous epidermal nevus.

DIFFERENTIAL DIAGNOSIS

1. Verrucous epidermal nevus
2. Nevus sebaceous

PATHOLOGICAL DISCUSSION

VEN is the most common type of hamartomatous keratinocytes proliferation. Post zygotic mutations in keratin 1 and / or keratin 10 have been hypothesized in its aetiology.¹⁰ Clinical variants documented by Blaschko are linear, zosteriform, unilateral or systematized patterns with streaks and swirls.^{6,11} It is known as systematized epidermal nevus, when it involves diffuse or extensive body areas^{12,13} and is called as nevus unius lateris, if it involves one half of the body.¹⁴ Ichthyosis hystrix is the term used to describe when there is extensive bilateral involvement.¹⁵

At the onset, lesions of VEN are slightly pigmented and becomes darker, verrucous and rough as the age progresses. It commonly involves trunk and limbs. Classically VEN are linear and vertical over limbs, transverse and S-shaped on trunk and normally do not cross midline. Until adolescence they may grow and remains unchanged afterwards.¹⁶ Face, head and neck region have been rarely involved,^{3,4,5} as in our case where involvement of face, neck, ears and scalp with scarring alopecia was seen.

Complications are uncommon and include maceration, infection of flexural lesions, recurrent paronychia if nail folds are involved. Rarely, neoplastic transformation into squamous cell carcinoma, basal cell carcinoma have been reported in few cases.^{17,18,19} Inflammatory linear verrucous epidermal nevus (ILVEN) also known as eczematous epidermal nevus, is a variant of VEN. It is clinically inflammatory in nature with psoriasiform histopathology. The onset of lesions is within the first six months of life and characterized by linear, erythematous, psoriasiform or eczematous papules associated with intense itching. Most commonly legs are involved, and it may extend to involve whole limb. In some cases, bilateral or systematized involvement have been reported.^{20,21}

Extensive nevi are frequently associated with other systemic abnormalities, especially of the eyes, central nervous system, musculoskeletal system and endocrinological system, then called as "epidermal nevus syndrome."^{22,23,24} Histologically, VEN are characterized by hyperkeratosis, acanthosis and papillomatosis with or without focal hypergranulosis and parakeratosis. Other less common histological variants reported are acrokeratosis verruciformis like, seborrheic keratosis like, verrucoid, psoriasiform, epidermolytic hyperkeratosis, focal

acantholytic dyskeratosis, porokeratosis-like and nevus comedonicus type.²⁵

In ILVEN, characteristic histological features seen are sharply demarcated columns of orthokeratosis and hypergranulosis alternating with parakeratosis and hypogranulosis with lympho-histiocytic infiltration in upper dermis.²⁶ VEN poses a serious cosmetic problem, especially if it is located on exposed parts. It is often difficult to treat, and results are variable and often disappointing. Various modalities of treatment have been tried till update.²⁷

This is a rare case of Systematized VEN with involvement of face, bilateral ears, neck, left side of back and scalp with scarring alopecia, causing lot of cosmetic disfigurement and mental trauma to patient and her parents. As there are chances of malignant transformation, long term follow-up is necessary for early detection and initiation of treatment.

DISCUSSION OF MANAGEMENT

Topical therapies such as salicylic acid, lactic acid, retinoic acid, topical calcipotriol, 5-fluorouracil, bleomycin, podophyllin, chemical peels have been tried, but are of limited benefits. Electrofulguration, radiofrequency and cryotherapy can be used for small lesions, with variable results.²⁷ Lasers such as carbon dioxide vaporization²⁸ and erbium: ND YAG laser therapy²⁹ have shown some beneficial effects. A case of recalcitrant VEN successfully treated with photodynamic therapy have been reported.³⁰ Surgical excision with grafting is the better modality of treatment comparatively, but may not be possible when there is extensive involvement.

FINAL DIAGNOSIS

Systematized verrucous epidermal nevus.

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