

## Case Report

# Systematised verrucous epidermal naevus with bilateral involvement

## 雙側受累的系统性疣狀表皮痣個案

P Aggarwal, K Aggarwal, VK Jain

---

Verrucous epidermal naevi (VEN) are congenital malformations of the epidermis consisting of verrucoid scaly plaques on the skin, following the lines of Blaschko. Involvement can be localised or generalised as in systematised naevi. Herein, a case of systematised VEN with bilateral involvement is reported, which is a rare presentation.

疣狀表皮痣屬於表皮的先天性畸形，皮膚可見沿著布拉許口氏線分佈的疣狀鱗狀斑塊。這一類痣可以只累及局部皮膚，反之則是廣泛分佈的系统性痣。本文報告了一例臨床罕見的雙側受累的系统性疣狀表皮痣。

**Keywords:** Epidermal naevus, bilateral involvement, ichthyosis hystrix

**關鍵詞：**表皮痣、雙側受累、高起魚鱗癬

---

## Introduction

Epidermal naevi are a heterogenous group of benign congenital hamartomas that may be composed of cells of sebaceous, apocrine, eccrine, follicular, or keratinocyte origin. Verrucous epidermal naevi (VEN) are a type of epidermal

naevi of keratinocyte differentiation.<sup>1</sup> Although multiple lesions of VEN are often seen, lesions presenting bilaterally are rare. We present a case of VEN involving the trunk and extremities, bilaterally.

## Case report

A 7-year-old boy presented with a history of asymptomatic, brownish raised lesions that first appeared over back of trunk at around one year of age and subsequently involved the hands and feet, bilaterally, over six months. His developmental milestones were normal and past medical history was unremarkable. No prenatal complications were reported. There was no family history of congenital anomalies.

---

**Department of Dermatology, Venereology and Leprology, Pt. B.D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India**

P Aggarwal, MD

K Aggarwal, MD

VK Jain, MD

Correspondence to: Dr. P Aggarwal

H. No. 1775, Sector 10/A, Gurgaon-122001, Haryana, India

Cutaneous examination revealed multiple, hyperpigmented, verrucous papules coalescing to form well-demarcated plaques with irregular borders, distributed bilaterally over the back and buttocks, in a blaschkoid pattern, acquiring the form of asymmetric V-shaped and transverse narrow bands (Figures 1a, b). Similar skin lesions were present linearly over the dorsal aspects of bilateral hands and feet (Figures 1c, d). The hair, nails and oral mucosa were normal. Skeletal, ophthalmological, dental, ear, nose and throat and neurological consultations revealed no abnormalities.

Histopathological examination of skin biopsy demonstrated hyperkeratosis, papillomatosis and acanthosis with prolonged rete ridges (Figure 2).

The patient was diagnosed as non-epidermolytic bilateral systematised verrucous epidermal naevus. He was treated with multiple sessions of cryotherapy with good cosmetic results and

advised to follow up once every six months to check for any late onset systemic symptoms.

## Discussion

VEN are mosaic lesions, originating from post-zygotic mutations and reflecting the dorsoventral outgrowth of the mutant cells during early embryogenesis.<sup>1</sup> The involvement can be localised or generalised but always follows Blaschko's lines, which represent patterns of epidermal migration.<sup>2</sup>

They are seen in approximately 1 in 1000 live births,<sup>1</sup> and manifest as linear, hyperpigmented, verrucous papules and plaques, usually at birth or infancy. They may enlarge slowly during childhood and become stationary at puberty.<sup>2</sup>

Verrucous epidermal naevi can exist as single or multiple lesions. They are said to be systematised

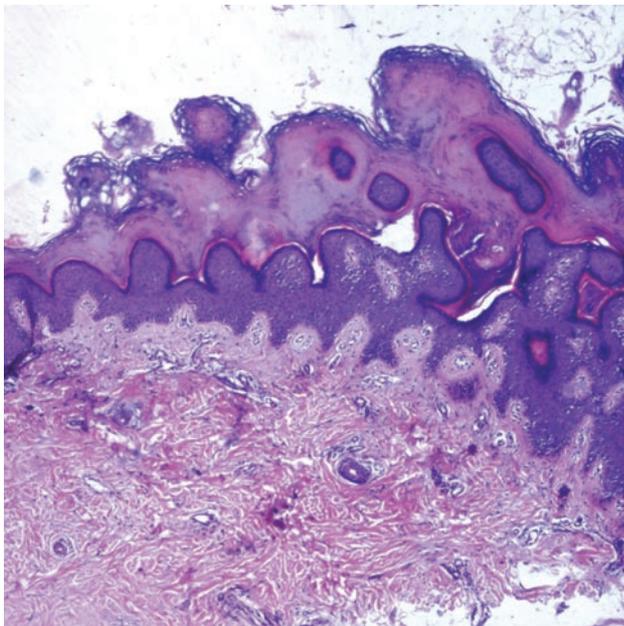


**Figure 1.** Hyperkeratotic papules arranged in streaks, distributed bilaterally over (a) back, (b) buttocks, (c) dorsum of hands and (d) feet.

when they occur as more than one linear lesion as in Lever's classification of epidermal naevi.<sup>3</sup> Systematised VEN usually involve one half of the body and hence are called, naevus unius lateris,<sup>4</sup> the term first used by von Baerensprung in 1863.<sup>5</sup> Bilateral involvement is rare and is also called ichthyosis hystrix.<sup>4</sup>

Histologically, VEN are subdivided into epidermolytic and non-epidermolytic types. It is important to distinguish the histological features of epidermolytic hyperkeratosis, in which case, patients may transmit the mutated gene to their offspring, resulting in a widespread cutaneous involvement.<sup>2</sup>

Around 30% of VEN, especially, in systematised cases, are often associated with multisystem abnormalities of the central nervous system, eyes and skeleton, as defined by the spectrum of epidermal naevus syndrome.<sup>3</sup> This warrants a thorough mucocutaneous, neurological, ophthalmological, and orthopaedic examination



**Figure 2.** Histopathology demonstrating hyperkeratosis, papillomatosis and acanthosis with prolonged rete ridges (H&E, 10X).

in patients with VEN, with specific investigations depending on the involved system. Also, lifelong close observation and follow-up is necessary, due to the risk of malignant transformation of the naevi and systemic manifestations which may sometimes present late in adulthood.<sup>3</sup>

In our case, despite having a systematised naevus, the patient had no evidence of any systemic involvement. The presence of multiple, bilateral epidermal naevi, suggests that the timing of mutation was during early embryonic development, causing the abnormal cells to be widely distributed.<sup>1</sup>

Management of VEN consists of medical and surgical treatment. Suggested medical treatments include dithranol, intralesional injection of corticosteroids, cryotherapy with liquid nitrogen, podophyllin, retinoids, topical calcitriol, topical 5-fluorouracil, and topical vitamin D3.<sup>5</sup> Definitive treatment involves full-thickness excision, which is impractical in extensive lesions. Carbon dioxide laser is an alternative option. Cryotherapy was used in our patient due to low cost and good cosmetic results in previous studies.<sup>6</sup>

## References

1. Shah KN, Honig PJ, Yan AC. Bilateral symmetric facial epidermal nevus. *J Am Acad Dermatol* 2007;56(Suppl):S51-3.
2. Brandling-Bennett HA, Morel KD. Epidermal nevi. *Pediatr Clin North Am* 2010;57:1177-98.
3. Dhanaraj M, Ramalingam M. Systematised epidermal nevus - a case report. *J Clin Diagn Res* 2015;9:WD01-2.
4. Mishra V, Saha A, Bandyopadhyay D, Das A. Bilateral Systematized Epidermolytic Verrucous Epidermal Nevus: A Rare Entity. *Indian J Dermatol* 2015;60:397-9.
5. Askar I, Aytakin S. Linear verrucous epidermal nevus with cutaneous horn. *J Eur Acad Dermatol Venereol* 2003;17:353-5.
6. Panagiotopoulos A, Chasapi V, Nikolaou V, Stavropoulos PG, Kafouros K, Petridis A, et al. Assessment of cryotherapy for the treatment of verrucous epidermal naevi. *Acta Derm Venereol* 2009;89:292-4.