

A case of zosteriform verrucous epidermal nevus at an unusual location

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Abstract Verrucous epidermal nevus is a common type of keratinocyte hamartoma present at birth or occurring later in life. It affects about one in every thousand live births. It is seen at any site, usually found on the lower extremities but is less common on the head and neck. We present a unique case of a female patient aged 12 years affected by zosteriform verrucous epidermal nevus over neck, a rare presentation.

Key words

Epidermal nevus, epidermal verrucous nevus, keratinocytes, zosteriform.

Introduction

Naevus is the Latin word for 'maternal impression' or 'birthmark' and indicates a circumscribed, non-neoplastic skin or mucosal lesion, usually present at or soon after birth, and fixed.

Verrucous epidermal nevi are congenital, noninflammatory cutaneous hamartomas composed of keratinocytes, each lesion comprising the progeny of a single mutant keratinocyte.

Case Report

A 12-year-old girl from rural area, presented to our department with the history of multiple asymptomatic skin lesions distributed over right side of the chest, shoulder, neck and upper back since birth. There was no history of delayed developmental milestones or seizure disorders, no consanguinity in parents, no

other children affected in family. On systemic examination, no abnormality was revealed. Following investigations of complete blood counts, ESR and urine routine examination were within normal limits.

The clinical examination revealed multiple, well-defined, irregular, brownish-black plaques, with warty surface, distributed between right C₂-C₄ areas and in segmental distribution but not crossing the midline (**Figures 1 and 2**). Hair, nail, and mucous membranes were normal.

A provisional diagnosis of verrucous epidermal nevus was made and a punch biopsy was done. Histopathology showed hyperkeratosis with irregular papillomatosis and acanthosis, presence of keratin plugs within the epidermis, and slight increase in basal melanin pigment. Dermis showed sebaceous glands proliferating vascular channels and very sparse lymphatic infiltrates, few eccrine glands with sparse melanin incontinence (**Figures 3 and 4**).

On the basis of clinical and histological evidence, a diagnosis of right sided zosteriform verrucous epidermal naevus of C₂-C₄ was made.

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Figure 1 Multiple, well-defined, brownish-black plaques, with warty surface, distributed between right C₂-C₄.



Figure 2 Warty plaques that never crossing midline of body.

Discussion

Verrucous epidermal nevus is a common type of keratinocyte hamartoma present at birth or occurring later in life. They affect about one in every thousand live births. It is seen at any site but is less common on the head and neck. Epidermal verrucous nevus is seen as vertical, linear or s-shaped lesion and does not normally cross midline.¹

Verrucous epidermal nevus follows the lines of Blaschko, suggesting that they represent postzygotic mutations. In general, larger

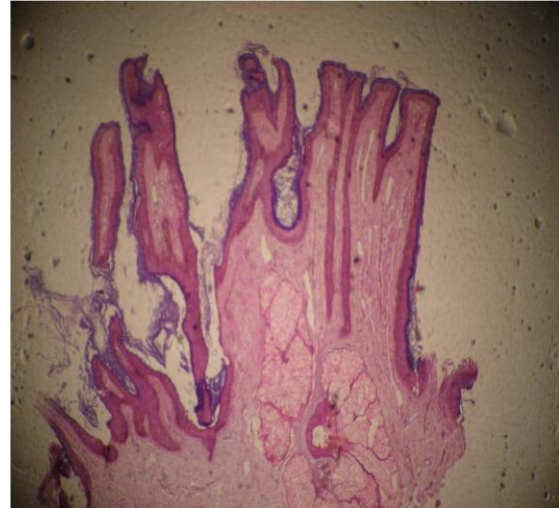


Figure 3 Hyperkeratosis, papillomatosis and acanthosis. Slight increase in basal melanin pigment is also present.

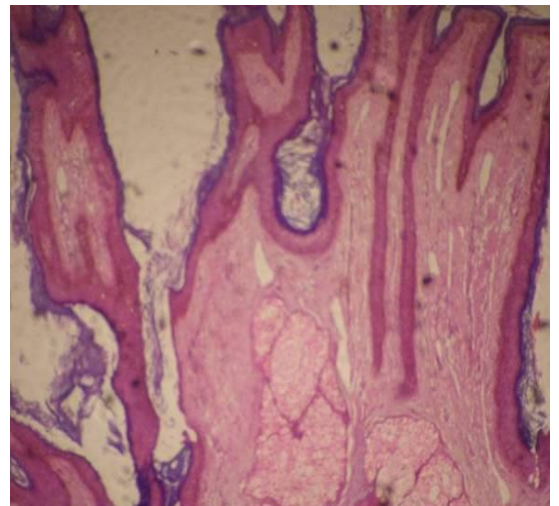


Figure 4 Dermis showed sebaceous glands, proliferating vascular channels and very sparse lymphatic infiltrates, few eccrine glands with sparse melanin incontinence.

lesions, more widespread lesions, and lesions of the head and neck are more likely to have associated internal complications. The combination of an epidermal nevus and an associated internal problem is called “epidermal nevus syndrome”.^{1,2}

Epidermal nevi most commonly present as a single linear lesion, but sometimes multiple unilateral or bilateral linear plaques are seen. Most lesions consist of well-circumscribed, hyperpigmented, papillomatous papules or plaques that are usually asymptomatic. Rarely, epidermal nevi are hypopigmented. Once

developed, the nevi may thicken and become more verrucous, especially over joints and in flexural areas such as the neck. Very rarely seen over shaft of penis,³ may be associated with features of woolly hair nevus,⁴ eyelid abnormalities causing trichiasis⁵ and may be seen as linear epidermal nevus over palm.⁶

Histopathology of epidermal nevus shows epidermal hyperplasia, hyperkeratosis, acanthosis, papillomatosis, and variable parakeratosis.

In our case, a girl aged 12 years presented with multiple, well-defined, irregular, brownish-black plaques, with warty surface, distributed between right C₂-C₄ areas and in segmental distribution. Histology showed hyperkeratosis with irregular papillomatosis and acanthosis, keratin plugs within the epidermis and slight increase in basal melanin pigment.

Based on clinical and histopathological examination diagnosis of zosteriform verrucous epidermal nevus was made, rare presentation over neck.

Treatment involves topical therapy such as corticosteroids, retinoic acid, tars, anthralin, 5-fluorouracil and podophyllin have all been used, but they are of limited benefit. Surgical modalities include excision with full thickness graft,⁷ cryotherapy,⁸ carbon dioxide laser vaporization⁹ and erbium:YAG laser therapy.¹⁰

Conclusion

Verrucous epidermal nevus is due to overgrowth of keratinocytes (horny skin cells). Lesions are present most commonly during birth or in first year of birth or early childhood. Our patient presented with multiple verrucous epidermal nevi seen in a zosteriform pattern with involvement of right C₂-C₄ dermatomes of neck, which is an uncommon presentation.

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