

## Epidermolytic hyperkeratosis in verrucous epidermal nevus

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### Abstract

Epidermal nevi are hamartomas that are characterized by hyperplasia of the epidermis and adnexal structures. Most cases arise sporadically during development of the embryo, but familial patterns of inheritance have been observed. Only 5-10% of patients with verrucous epidermal nevus show histological features of epidermolytic hyperkeratosis. This has a significant clinical importance as this patient carry the risk of parenting a child of congenital bullous ichthyosiform erythroderma. A 7-year-old boy born of non-consanguineous marriage presented with non-pruritic multiple verrucous papules that coalesce to form well-demarcated, skin colored to brown, papillomatous plaques over the right side of abdomen, right loin, right legs including a cauliflower like verrucous plaque over the umbilicus distributed along the line of Blaschko noticed since 1-year of age. Oral cavity, palms and soles were spared. Developmental milestones were normal. A diagnosis of verrucous epidermal nevus was made. Histology showed hyperkeratosis, papillomatosis and vacuolization of the cells in the stratum spinosum and granulosum with keratohyalin granules. Routine examination of blood and chest X-ray were normal. This case is being reported to highlight the rare histological features and to re-emphasize the importance of routine histological examination and genetic counseling in all cases of verrucous epidermal nevus.

**Key Words:** Congenital bullous ichthyosiform erythroderma, Epidermal nevus, Epidermolytic hyperkeratosis

### INTRODUCTION

There have been case reports of epidermal nevi showing histological features of epidermolytic hyperkeratosis (EH) in the past. More recently, this association has been found to be of significant clinical importance.

EH is a pathologic reaction pattern of skin. This pattern has been observed incidentally in a variety of benign and malignant skin lesions or hereditary disorders.<sup>[1]</sup>

Epidermal nevi are hamartomas that are characterized by hyperplasia of the epidermis and adnexal structures. Only 5-10% of patients with verrucous epidermal nevus show histological features of EH.<sup>[1]</sup> This has a significant clinical importance as this patient carry the risk of parenting a child of congenital bullous ichthyosiform erythroderma (CBIE).<sup>[2]</sup>

### CASE REPORT

A 7-year-old boy born of non-consanguineous marriage presented with non-pruritic multiple verrucous papules that

coalesce to form well-demarcated, skin colored to brown, papillomatous plaques over the right side of abdomen, right loin, right legs including a cauliflower like verrucous plaque over the umbilicus distributed along the line of Blaschko [Figures 1 and 2] noticed since 1-year of age. Oral cavity, palms and soles were spared. Developmental milestones were normal. There was no significant medical or family history. Systemic review was otherwise normal. A skin biopsy showed a hyperplastic epidermis. There was alternating hyperkeratosis and parakeratosis as well as vacuolar degeneration. Some epidermal cells were hypereosinophilic. In addition, coarse keratohyalin granules were seen in the granular layer [Figure 3]. All these were consistent with EH. Routine examination of blood and chest X-ray were normal.

Patient was treated with 10 mgs of isotretinoin daily along with topical keratolytic agents and the patient was managed with regular follow-up.

### DISCUSSION

Epidermal nevi are common hamartomatous lesions arising from embryonic ectoderm affecting about 1 in 1000 people.<sup>[2]</sup> Blaschko documented that linear streaks and swirls adopted by these nevi may be due to somatic mutation, each lesion comprising the progeny of a single mutant keratinocyte.<sup>[3]</sup> In some cases, most commonly when there are multiple lesions, there may be associated defects in other tissues, particularly the skeleton and the

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**Figure 1:** Papillomatous plaques over the right side of abdomen, right loin including a cauliflower like verrucous plaque over the umbilicus distributed along the line of Blaschko



**Figure 2:** Close up picture showing the verrucous lesions



**Figure 3:** Photomicrograph demonstrating hyperplastic epidermis with alternating hyperkeratosis and parakeratosis. Coarse keratohyalin granules are also seen in the granular layer

central nervous system, here the term “epidermal nevus syndrome” is often applied.<sup>[4]</sup>

Epidermal nevi tend to follow the lines of Blaschko. The disease may be a mosaic disorder resulting from a postzygotic mutation.<sup>[2]</sup>

These nevi occur mainly on the trunk and limbs, although occasionally the head is involved, particularly as part of a very extensive lesion. They may be congenital, but in over 50% of cases the onset is after birth, usually in the 1<sup>st</sup> year of life, but occasionally as late as adolescence.<sup>[5]</sup>

A rare subgroup of epidermal nevi is clinically indistinguishable from other epidermal nevi, but displays histopathological features typical of EH. This form of epidermal nevus arises from a postzygotic KRT1 (keratin) or KRT10 mutation in a cell destined to become an epidermal keratinocyte.<sup>[6]</sup> Patients with this type of epidermal nevi sometimes have offspring with generalized EH more specifically the variant of CBIE, which is characterized by mechanical stress-associated skin blistering arising from fragile cells and rupturing within the suprabasal layers of the epidermis.<sup>[2]</sup>

## CONCLUSION

Ideally, all linear epidermal nevi (especially if extensive) should be biopsied. If EH is present, the parents should be given genetic counseling and offered prenatal diagnosis. In the future, deoxyribonucleic acid sequencing techniques will permit prenatal diagnosis at a much earlier gestational age.

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