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Keratinocytic Verrucous Epidermal Nevi in a Child

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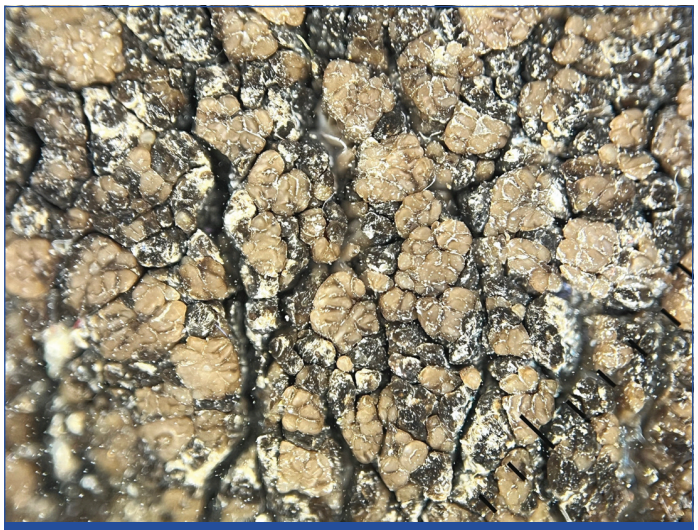
A four-year-old male child was brought by the parents to the Dermatology Outpatient Department (OPD) with the complaint of a unilateral asymptomatic dark-coloured lesion over the right-side of the trunk, back, and upper limb since birth. The parents reported that the lesions increased in size proportionate to body growth, and the surface has become darker brown and rough. The child had age-appropriate weight and height and no history of neuro-developmental delay. Examination revealed large, dark brown, warty plaques in a linear pattern, strictly confined to the right half of the trunk and right upper limb. Two plaques (A and C) were seen to be extending from the anterior midline to the posterior midline [Table/Fig-1]. The other two plaques (B and D) were limited to only the posterior part of the right half of the trunk [Table/Fig-2].

[Table/Fig-1]: Clinical image showing warty-appearing hyperpigmented plaques from the right anterior midline extending to the posterior trunk and right arm.
[Table/Fig-2]: Clinical image showing four hyperpigmented verrucous plaques. (B and D) were limited to only the posterior part. (Images from left to right)

Contact polarised dermatoscopy using DermLite® DL4 showed multiple dark-coloured, closely set warty islands with fissuring in between, reminiscing of 'cauliflower appearance' [Table/Fig-3]. Serial excision was advised to the patient; however, due to poor socioeconomic conditions, they could not afford to seek the treatment.

Verrucous Epidermal Nevus (VEN) is a benign, non inflammatory epidermal nevus that usually appears at birth or within the first few years of life. Generally, affecting the trunk or extremities, it can arise anywhere on the body. VEN represents a clone of epidermal cells carrying a somatic mutation that arose early in post-zygotic embryogenesis [1].

When two genetically different cell populations arise from a post-zygotic mutation within an individual, it is referred to as somatic mosaicism, affecting a portion of the somatic cells in a particular body region [2,3]. Epidermal nevi can be caused by mutations in several different genes, including Rat sarcoma (RAS), Fibroblast Growth Factor Receptor 3 (FGFR3), phosphatidylinositol-4,5-bisphosphate 3-kinase catalytic subunit alpha (PIK3CA), and keratins (KTR1 and KRT10) [4,5]. Post-zygotic somatic mutations in the FGFR3 gene account for nearly 33% of VEN [6].



[Table/Fig-3]: Dermoscopy (10x magnification) showing dark-coloured closely set warty islands with fissuring in between reminiscing of 'cauliflower appearance'.

The nevi are due to genetic mosaicism, which reflects the migration paths of individual clones of genetically identical cells represented by Blaschko's lines. According to the various patterns, the present case fits in the lateralisation pattern of Blaschko's lines (type 5) [3].

Saini S et al., discussed five cases of biopsy-proven VEN ranging from three years to 21 years of age, with the most common presentation being on the trunk as discrete to closely arranged verrucous hyperpigmented plaques. The family history was positive in three cases. Histopathology showed hyperkeratosis, parakeratosis, acanthosis, orthokeratosis and papillomatosis [1].

Various treatment modalities have been used, including topical agents, cryotherapy, lasers, electrofulguration, and chemical peels, with varying clinical outcomes [6]. Future therapeutic options include targeted therapy, acting on various mutated genes/proteins, thereby downregulating the constantly active receptor/protein and thus, normalising the differentiation and proliferation of epidermal cells.

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