A 1.5-year-old Egyptian boy presented with widespread, brownish, hyperkeratotic, slightly verrucous plaques since he was two months of age. There was no past history of blistering or generalized cutaneous redness at or after birth, he had no medical problems and he showed normal growth and development. No other family members were affected by a similar skin condition.

Upon examination, brownish, verrucous plaques and papules in Blaschkoid distribution were found on the trunk and were more apparent on the extremities bilaterally (Figure 1A) and posterior of the neck (Figure 1B), as well as in the intertriginous areas. His hair and nails were normal, and general examination for other organ systems was unremarkable, the clinical differentials included linear psoriasis and lichen striatus.

A written informed consent was obtained from the parents of the boy and two punch skin biopsies from the lesions on his nape and knee were taken and their microscopic examination (Figure 1C, Figure 1D) revealed typical features of epidermolytic hyperkeratosis.

Epidermolytic hyperkeratosis (EHK) is one of the minor pathologic reaction patterns of the skin, first described by Ackerman in 1970, and characterized by hyperkeratosis, hypergranulosis, and epidermolysis\(^1\). Epidermolytic hyperkeratosis is a rare occurrence, by using the light microscopy, epidermolysis is seen as variable sized clear empty spaces around keratinocyte nuclei with indistinct cell boundaries and premature excessive formation of kerato-hyaline granules in the upper spinous and granular layers\(^2\).

The use of the term EHK to describe the characteristic histologic features of this disorder caused a lot of confusion in the literature. It has been described as the fundamental histopathologic feature of bullous congenital ichthyosiform erythroderma and as well as an incidental finding in other cutaneous disorders including melanocytic nevi, basal cell carcinoma, isthmus-catagen cyst, leukoplakia, epidermolytic acanthoma, as well as in normal skin and in normal mucosa of the oral cavity\(^3\). With all mentioned clinical sittings that EHK can be found in the pathology the final diagnosis of this case was very confusing and it cannot be reached unless suitable clinical data are correlated with the pathological findings. So; after constellation of the clinical and pathologic data a diagnosis of bilateral systematized verrucous epidermal nevus with epidermolytic hyperkeratosis was made.

Verrucous epidermal nevus (VEN), is a congenital, non-inflammatory cutaneous keratinocyte hamartoma. It is manifested as papillomatous papules or plaques, often linear or Blaschkoid in shape. The term systematized epidermal nevus is used for lesions that are bilateral and excessive, also known as nevoid ichthyosis hystrix\(^4\). Histologically, VEN show hyperkeratosis, hypergranulosis, acanthosis, and papillomatosis\(^5\). The difficulty in pathological diagnosis related to that there are at least 10 different pathologic patterns of epidermal nevi have been described, with any lesion possibly revealing more than one histological pattern\(^6\). EHK is one of these uncommon pathologic presentations of VEN (our case) it is found in only 16% of this clinical condition\(^7\). Few similar cases
were reported in the literature\textsuperscript{6,8,9}, so it is important for both the dermatologist and the pathologist to be aware of such interesting case.

References

Figure 1. Epidermolytic hyperkeratosis. Dark-brown verrucous plaques in the extensor of the knee (A) and posterior neck (B). Histopathology of the lesions showing: hyperkeratosis, hypergranulosis, acanthosis, papillomatosis and epidermolytic hyperkeratosis (hematoxylin-eosin, ×100) (original magnification) (C) and epidermolysis is seen as various sized clear spaces around keratinocyte nuclei with premature excessive formation of kerato-hyaline granules in the upper spinous and granular layers (hematoxylin-eosin, ×400) (original magnification) (D).