Quiz

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A six years old female is presented with lesion in the left upper limb from the shoulder to the nail. The lesions were verruciform, linear start to develop soon after birth (Fig 1). Biopsy showed orthokeratotic hyperkeratosis, moderate acanthosis and irregular papillomatosis (fig2 and 3).

Diagnosis: This lesion was diagnosed as linear verrcous epidermal Nevus.

Discussion:

Epidermal Nevi is a congenital hamaromas of embryonal ectodermal origin, classified on the basis of their main component. The component may be sebaceous, apocrine, eccrine, follicular or keratinocytic. An estimated 1/3 of individuals with EN have involvement to other organ systems. Linear verrcous epidermal Nevus is distinct from psoriasis. However, they may share some common pathogenic pathways. These pathways are probably mediated by interleukin 1, interleukin 6, tumor necrosis factor-alpha, and intercellular adhesion molecule-1.

ILVEN is a linear, persistent, pruritic plaque, usually first noted on a limb in early childhood. Originally described by Unna in 1896, a few patients were reported prior to 1971 when Altman and Mehregan3 delineated ILVEN as a distinct entity in 25 patients. They coined the name inflammatory linear verrucous epidermal nevus, labeling it a clinical and histopathologic type of linear verrucous nevus that is often inflammatory or psoriasiform. ILVEN accounts for approximately 5% of patients with EN and has been described in a mother and daughter. Six different syndromes with EN as part of them have been delineated. These include [1] Proteus, [2] congenital hemidysplasia with ichthyosiform nevus and limb defect, [3] phakomatosis pigmentokeratotica, [4] sebaceous nevus, [5] Becker nevus, and [6] nevus comedonicus[4] syndromes. The spectrum has recently been expanded with the description of linear Cowden nevus as a new distinct epidermal nevus.[5] This nonorganoid epidermal nevus is probably due to loss of heterozygosity, occurring at an early developmental stage in an embryo with a germline PTEN mutation, giving rise to Cowden disease.

References:

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7- *Rebollo Alvarez-Amandi M; Berciano BJ; Combarros PO; Figols Ladronde Guevara J: Jadassohn's nevus sebaceous and nevus unius lateris. Report of an observation with multiple dysplasia. Med. Clin. (Barc) 1979; 72: 106 – 110.*

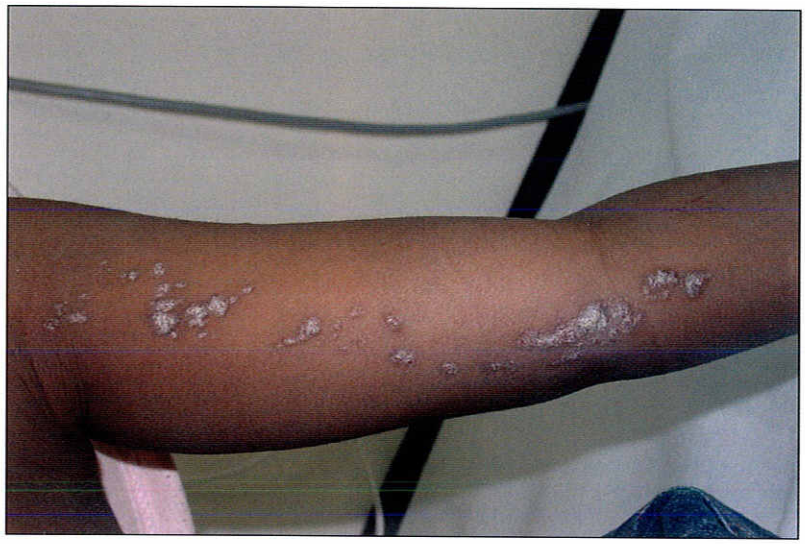

Fig 1

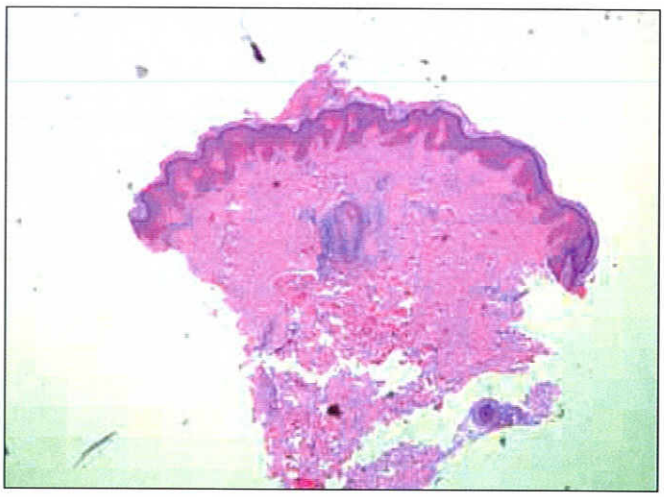

Fig 2

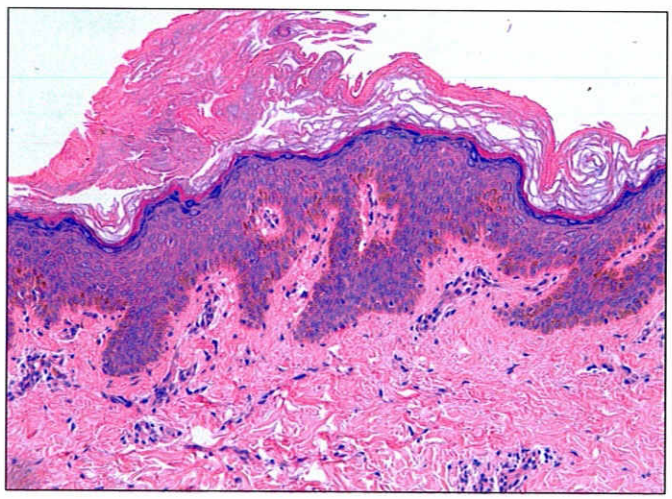

Fig 3