Organoid Nevus

Patient: M.K., a 7-year-old girl
Duration: Since birth
Distribution: Right side of the body including face, scalp, trunk, axilla, and right arm.

History: Asymptomatic skin lesions since birth. Patient is in good general health, no central nervous system or skeletal anomalies. Negative family history.

Physical Exam: Verrucous brown rough papules and plaques, with spiny projections and comedo like structures, distributed in a linear fashion. On the scalp are patches of alopecia with a yellowish hue.

Histopathology:
Trunk: Mild focal hyperkeratosis overlying the hair follicle, associated with mild papillomatosis and irregular hyperplasia. Scalp: Cystic dilatation of the middle and deep portions of the hair follicle with trichilemmal keratinization and abortive hair papilla arising from these dilatations. The findings are consistent with the organoid nevus.

Laboratory: None

Treatment:
Trial of Daivonex® ointment on one area, and Diprosalic® ointment on another.

Discussion:
Epidermal nevi are hamartomatous lesions arising in the embryonic ectoderm. Most epidermal nevi comprise more than one cell type and therefore the term organoid nevus is preferred. In some cases, most commonly when there are multiple lesions, there may be associated defects in other tissues, particularly the CNS and the skeletal system. The term epidermal nevus syndrome is applied in such cases. A genetic influence on the development of epidermal nevi is suggested. The linear streaks and swirls adopted by the hamartomas reflect mosaicism. A possible role for the dermis in their development is suggested by the difficulty experienced in ablating such lesions surgically without destruction of the underlying dermis.
• Organoid nevi are classified according to the predominant cell type:
  Keratinocyte - Follicular - Sebaceous - Apocrine - Eccrine Nevi
• Keratinocyte nevi include:
  Verrucous Epidermal Nevus - Acantholytic Epidermal Nevus - Nevus Corniculatus - Linear porokeratosis - Inflammatory Epidermal Nevus
**Verrucous Epidermal Nevi** are circumscribed hamartomatous lesions comprised almost exclusively of keratinocytes that tend to become more pigmented and verrucous over the years. Extension generally ceases by the end of adolescence after which lesions remain stable. They have been associated with scalp woolly hair and megalopinna when the ear is involved. Also they are associated with McCune-Albright and Klippel-Trenaunay syndromes.

**Acantholytic Epidermal Nevus** Darier-like appearing after the age of twenty and Hailey-Hailey-like epidermal nevus characterized by periods of spontaneous improvement followed by relapse.

**Nevus Corniculatus** featuring filiform and horn-like keratosis and giant comedones.

**Linear porokeratosis** reflecting mosaicism for the gene responsible for porokeratosis.

**Inflammatory Epidermal Nevi** include dermatitic, lichenoid, as well as linear psoriasis variants.

**References:**