ATROPHODERMA VERMICULATUM – A CASE REPORT

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ABSTRACT
We report a case of a 4 year old boy who had asymptomatic multiple pits over both his cheeks. A clinical & histopathological diagnosis of Atrophoderma Vermiculatum was made. It is presented for its rarity.

INTRODUCTION
Atrophoderma vermiculatum a rare condition, commonly seen in children, is a benign disorder of the hair follicles. It manifests as honeycomb or symmetric reticular atrophy of the cheeks, which may extend to forehead and ears. An abnormal keratinization in the pilosebaceous follicle was the proposed defect associated with this disorder. Management of atrophoderma vermiculatum is challenging.

CASE REPORT:
A 4-year old boy was brought by his parents with complaints of asymptomatic multiple depressed scars on both his cheeks since birth (Fig.1). The scars were preceded by multiple small raised papules. There was no history of photosensitivity or any other systemic complaints. There were no similar complaints in the family. On examination, symmetrical, closely crowded areas of atrophy, appearing as pits 1-3mm in size, separated by narrow ridges, producing a honeycomb pattern was noted over the cheeks confined between nasolabial folds and preauricular region. Milia and comedones were not present. The cheeks appeared shiny, waxy & firm on palpation. Scalp, palms, soles, nails, hair & oral cavity were normal. Systemic examination & other routine investigations were found to be normal. A 2.5mm punch biopsy was taken & histology revealed epidermal atrophy & enlarged, dilated hair follicles under low power. Dilated capillaries with infiltrates and edematous collagen were noted under high power (Figs. 2 and 3). Thus, a diagnosis of Atrophoderma Vermiculatum was made.

DISCUSSION:
Many synonyms such as Atrophoderma reticulatum symmetrica faciei; Ulerythema aceneiform; Atrophoderma reticulatum; Atrophodermia reticulata; Atrophodermia vermiculatum; Atrophoderma vermiculata; Atrophodermie vermiculée des joues avec kératoses folliculaires; Honeycomb atrophy; Folliculitis ulerythema reticulata; Folliculitis ulerythemosa Folliculitis ulerythematous reticulata have been used for this condition. They are a reflection on the uncertain nosologic status of this rare clinical entity. “Ulerythema” means scaring & redness; “vermiculatum” means worm-eaten.

Atrophoderma vermiculatum is inherited as an autosomal dominant disorder. It has been reported in a father and son by one of the authors (Thomas J, Muthuswami TC, 1987).
Both sexes are equally affected (Frosch et al., 1988). The atrophic lesions of the cheek may be preceded by multiple symmetrical inflammatory papules, which become pitted, atrophic and form depressed scars. These scars form a reticulated or honeycomb pattern. Erythema & comedones may be present depending upon the degree of follicular plugging. The lesions can extend to the pre auricular region or forehead. It usually occurs during childhood & it’s of a slow progressive course. It can occur rarely unilaterally (Arrieta E, Milgram-Sternberg Y, 1988). The pathogenesis of atrophoderma vermiculatum is abnormal follicular hyperkeratinization of upper third of the hair follicle, leading to obstruction of the growing hair shaft and production of chronic inflammation. The net result is scarring below the level of obstruction. Biopsy reveals atrophied epidermis, dilated capillaries with perivascular round cell infiltrate. Basophilic degeneration with edematous collagen may also be seen. Hair follicles appear to be dilated, tortuous & hyperkeratotic. Horn cysts may be seen occasionally. Atrophia maculosa varioliformis cutis, keratosis pilaris rubra of the cheeks & erythromelanosis faciei are the differential diagnosis to be considered.

Atrophoderma vermiculata belongs to a group of related diseases, ‘keratosis pilaris atrophicans’, which also includes keratosis follicularis spinulosa decalvans and ulerythema ophryogenes. These 3 conditions are characterized by follicular keratotic papules, variable degrees of inflammation & secondary atrophic scarring. Ulerythema ophryogenes also called as keratosis pilaris atrophicans faciei clinically differs from atrophoderma vermiculata by involving the lateral portion of the eyebrows & progression ceases after puberty. Its mode of inheritance is autosomal dominant trait with incomplete penetrance. Keratosis follicularis spinulosa decalvans involves eyebrows, scalp & extremities with scarring alopecia. It is of X- linked recessive inheritance.

Various syndromes associated with atrophoderma vermiculata include Rombo syndrome (milia, telangiectasias, basal cell carcinoma Hypotrichosis, acral cyanosis & rarely trichoepitheliomas) (Van Steensel et al., 2011). Nicolau-Balus syndrome (syringomas & milia), Tuzun syndrome (scrotal tongue) & Braun–Falco–Marghescu syndrome (keratosis pilaris & palmoplantar hyperkeratosis).

Treatment modalities include topical and systemic medication as mentioned below:
Topical: emollients, tretinoin, corticosteroids and keratolytics have been tried but are ineffective.
Systemic isotretinoin has shown some improvement, by preventing the progression of atrophy and inducing remission (Callaway SR, Lesher JL, 2004). Laser therapy with CO2 and 585nm pulse has been reported to be effective (Hidetoshi Takahashi et al., 2012). Dermabrasion & fillers such as collagen, hyaluronic acid & autologous fat are also being tried.

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**Figure 1:** clinical picture showing multiple pitted, atrophic and form depressed scars in a reticulated or honeycomb pattern over Bilateral cheeks.

**Figure 2:** Histological picture (low power- 100 X), showing epidermal atrophy & enlarged, dilated hair follicles

**Figure 3:** Histological picture (high power- 400 X), showing Dilated capillaries in the dermal papillae with infiltrates and edematous collagen
REFERENCES