Nevus comedonicus in a Filipino girl

Nevus comedonicus is a rare developmental anomaly of the pilosebaceous unit presenting as grouped dilated follicular orifices plugged with keratin. The comedones are usually arranged unilaterally along the lines of Blaschko. As of year 2000, only 200 cases have been reported. (1)

We document a case of nevus comedonicus in an 8-year-old Filipino girl, who presented with increasing number of discrete papules and comedones on her trunk, both axilla and posterior thighs for a period of 1 year.

Cutaneous examination revealed multiple, discrete skin-colored papules some closed, some with dilated follicular openings with firm grayish to black hyperkeratotic plugs, measuring 0.1 x 0.1 cm to 0.2 x 0.2 cm over the right and left axilla, chest, abdomen, posterior trunk and medial and both posterior thighs, nasal bridge, and left cheek.

The patient was managed with topical glycolic acid cream and retinoids. Gentle extraction of the horny plugs in the open comedones was done as well.

Nevus comedonicus was first described by Kofmann in 1895 as *comedo nevus*, other synonyms include *nevus acneiformis unilateralis, nevus folliculoris keratosis* and *nevus zoniforme*. Lesions are usually present at birth or develop during the first 2 decades of life. It is a non-familial disorder and tends to occur sporadically. It becomes designated as Nevus Comedonicus Syndrome when lesions occur in combination with other development anomalies such as that of the central nervous, skeletal, and ophthalmologic systems. Rarely, benign or malignant tumors may arise over nevus comedonicus lesions such as trichofollicloma, pilar sheath acanthoma, hidradenoma papilliferum, syringocystadenoma, and squamous cell carcinoma. [2]

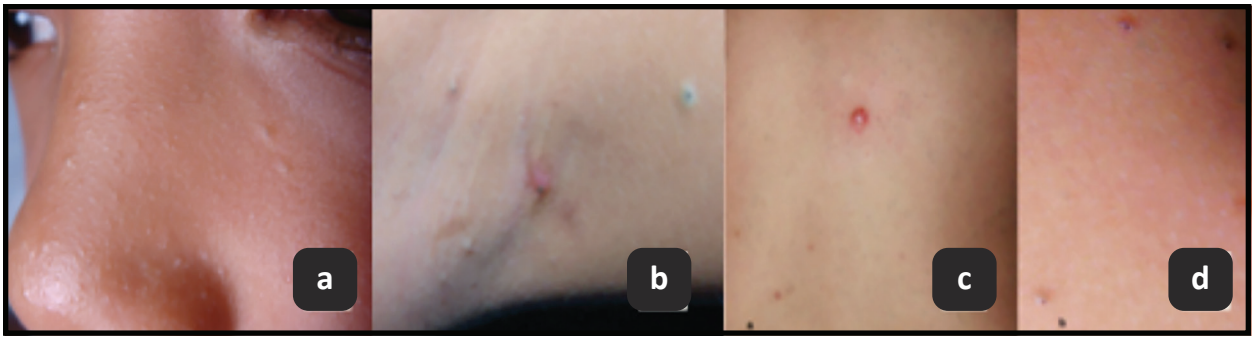

Figure 1. (a-d) Multiple discrete comedonal papules on the face, axilla, chest and thighs.

No other organ system involvement was noted upon further examination.

Routine hematologic, urinary, biochemical and radiologic findings were all normal. A 4- mm skin punch biopsy of a comedonal lesion on the left axilla revealed widely dilated infundibulum that opens directly into the epidermis with keratinous material inside the follicle. The dermis was unremarkable with no sign of inflammation.

It has been postulated that it is a hamartoma of the pilosebacous unit, with failure of mesodermal part to differentiate properly producing only soft keratin, which accumulates in the adnexal orifices presenting as comedone-like lesions.[1]

Differential diagnosis for nevus comedonicus are familial dyskeratotic comedones, trichostasis spinulosa, eruptive vellus hair cyst and comedonal acne. Histopathologically, both trichostasis spinulosa and eruptive vellus hair cyst will show obstructed follicular infundibula with vellus hair, while the absence of inflammatory infiltrates will differentiate it from comedonal acne. Nevus comedonicus will show

deep, wide invaginations of acanthotic epidermis filled with concentric lamellae of keratin with rudimentary hair follicular structures, and sebaceous glands.[2,3]

Treatment and management of nevus comedonicus have shown varying and limited successes. The main objective in its management is to control the formation of comedones and prevent the progression of lesions into an inflammatory type. Non-inflammatory lesions are managed with comedone extraction, use of pore strips, tretinoin, ammonium lactate, tazarotene and calcipotriene. Inflammatory lesions would require antibiotics, oral retinoids and/or surgical excision.[1,2] Complications of recurrent inflammation may lead to fistula formation and massive disfigurement.

Case reports on the use of ultrapulse CO2 laser, erbium-YAG lasers[3] repeated microneedling[4] and microneedling with radiofrequency[5] have shown more promising long-term results as part of the emerging management of nevus comedonicus.

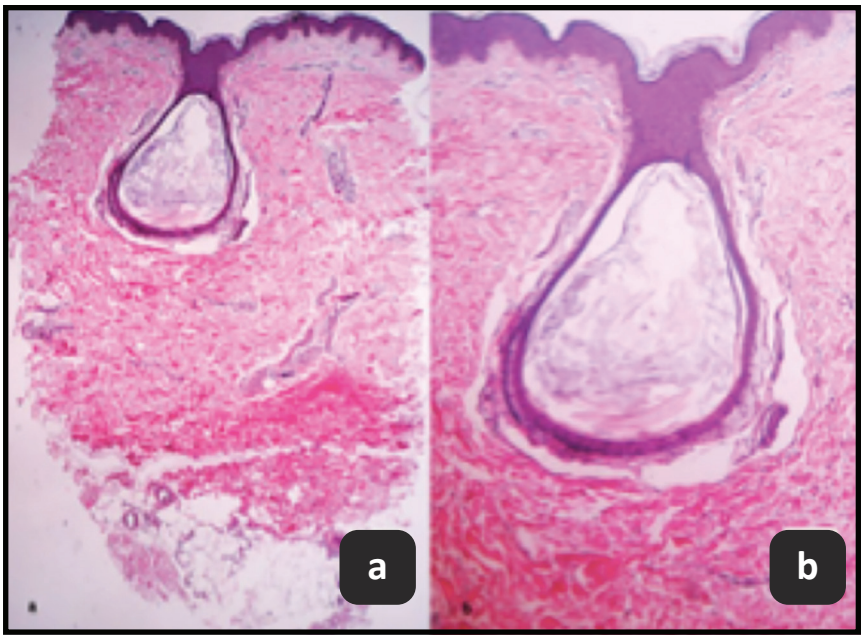

Figure 2. Biopsy specimen from the L axilla. a.) Low magnification shows a single widely dilated follicular infundibulum that contains keratinous material that does not open into the epidermis. b.) On higher magnification, no inflammatory infiltrates in the dermis can be seen. H & E x 40 (a), x 100 (b)

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