



NEVUS COMEDONICUS FOLLOWING BLASCHKO'S LINE- A RARE CASE REPORT

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ABSTRACT

Nevus comedonicus is a rare adnexal hamartoma of pilosebaceous unit of unknown etiology characterized by closely arranged dilated follicular opening with keratinous plugs resembling classical comedones¹. It may be a part of nevus comedonicus syndrome characterized by ocular, skeletal and central nervous system abnormalities. Seven month old female child presented with group of pits filled with keratinous plugs along Blaschko's line resembling comedones without any systemic abnormalities³.

KEYWORDS : Nevus comedonicus, Nevus comedonicus syndrome, Blaschko's line.

INTRODUCTION:

Nevus comedonicus is an uncommon skin abnormality of unknown etiology, first described by Koffman in 1895. Fewer than 200 cases having been reported. It is characterized by an aggregation of dilated follicular orifice filled with keratinous material.

CASE REPORT

Seven month old female child born of non- consanguineous marriage presented with asymptomatic group of pits filled with dark coloured keratinous plugs resembling comedones over left axilla extending up to elbow in a linear fashion since birth. Her mother also gives history of development of few pustules and abscess with purulent discharge since one month. Her developmental milestones were normal and there was no history of seizures or similar lesions in the family members.

General and systemic examination including eye, skeletal and central nervous system were normal. Dermatological examination revealed multiple grouped comedones over the left axilla (Fig.1) and extending up to elbow in a linear fashion following Blaschko's line (Fig.2). Few pustules and an abscess with purulent discharge were seen (Fig.3).



Fig 1: Grouped comedons

Fig 2: Comedones along blaschko's line

Fig 3: Pustule & abscess

Skin biopsy of the lesion showed hyperkeratosis and acanthosis (Fig.4). Epidermis show deep & wide invagination resembling dilated hair follicle. It is filled with horny keratinous material penetrating the reticular dermis (Fig.5). Few immature hair follicles were seen.

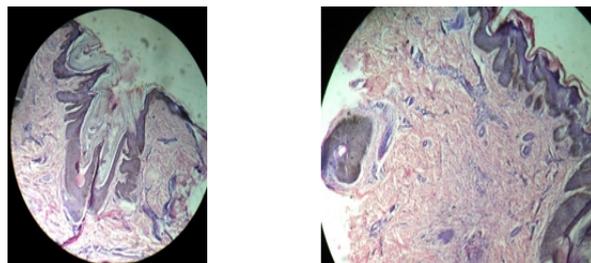


Fig 4: Keratin filled dilated epidermal invagination

Fig 5: Epidermolytic HK with immature hair follicle

Hence, the patient was diagnosed with an inflammatory variant of nevus comedonicus. She was managed with topical and systemic antibiotic and her mother was reassured.

DISCUSSION:

Nevus comedonicus is a rare skin disorder. Clinically it is characterized by closely arranged, dilated follicular openings with keratinous plugs resembling classical comedones. The disease was first described in 1895 by Kofmann¹ who suggested the term 'Comedo nevus'. It is of unknown etiology. Recently it was proposed that overstimulation of FGFR2 signaling with increased expression of interleukin-1 α may be involved in Nevus comedonicus pathogenesis². Somatic mutations like Ser252Trp substitution have been identified in individual patients with segmental acne³

Nevus comedonicus can occur congenitally or develop later in life, most commonly at the age of approximately 10 years. Nevus comedonicus is part of the Nevus comedonicus syndrome, a term coined by Engber⁴ in 1978.

They can appear anywhere on the body but are most commonly found on the face, trunk, neck and upper extremities and rarely other anatomical regions like genital area, palms, and soles. The lesions appear as closely grouped slightly elevated papules with central keratinous plugs giving the appearance of comedones. They may be distributed in a linear, interrupted, segmental, unilateral or bilateral pattern and sometimes follow the lines of Blaschko.

Two types of nevus comedonicus have been identified; the first is non-pyogenic variant characterized by comedone like lesions and a second type, inflammatory variant characterized by formation of cysts, papules, pustules, and abscesses in various stages of development.

The major histopathological features are large grouped, dilated follicular ostia devoid of hair shafts but filled with keratin layers. At some locations in the bases of the follicular invaginations there may be rudimentary glands. Sometimes large cysts and immature hair follicle may be seen. Hyperkeratosis (epidermolytic hyperkeratosis) and acanthosis of the epidermis may be present.

Nevus comedonicus syndrome (ORPHA64754) is a neurocutaneous disorder which consists of nevus comedonicus with skeletal, ocular, and central nervous system abnormalities. The most common symptoms include cataracts, scoliosis, fused vertebrae, spina bifida, and delayed mental development.⁵ In addition, seizures, paresis,

dysgenesis of corpus callosum, electroencephalographic abnormalities, limb deformities such as absence of the fifth finger, polysyndactyly or clinodactyly, oligodontia, and other skin disorders such as ichthyosis, leukoderma, lichen striatus, linear morphea, Sturge-Weber syndrome, hemangiomas have been reported.

Nevus comedonicus should be differentiated from acne vulgaris, acne neonatorum, milia, nevus sebaceous and linear Darier disease. Non inflammatory variant is asymptomatic and treatment is based on cosmetic concern. Inflammatory variant is treated with topical retinoids,⁷ tacrolimus, calcipotriene⁸ and intralesional steroids to control inflammation. Keratolytics may be of use. Systemic antibiotics may be used to control infection. Surgical excision, extraction, dermabrasion and laser resurfacing can also be useful cosmetically.

Laser treatments with 2,940-nm Erbium YAG, 10,600-nm Ultra pulsed CO₂, or 1,450-nm Diode lasers have shown improvement in Nevus comedonicus patients.⁹

Erbium YAG¹⁰ laser is known to cause superficial ablation but in nevus comedonicus the epidermal invagination is up to papillary dermis so, Erbium YAG laser treatment is often followed by delayed relapses.

Ultra pulsed CO₂¹¹ have an advantage over Erbium YAG laser with more ablative depth. With ultra pulse CO₂ laser, the energy spreading down the hair follicle produced cavitation and a zone of collagen damage 300 to 400 µm down the follicle wall and gives good intra operative hemostasis. This has been associated with excellent tissue healing with minimal thermal damage as the ultra pulse mode has a tissue thermal impact less than the thermal relaxation time of skin. Ultra pulse CO₂ laser has been proven to have a specific follicular penetration.

In contrast to ablative lasers, the 1,450 nm Diode penetrates up to papillary dermis induces thermal injury up to 400 to 500 µm in and around the sebaceous gland that results in shrinkage of sebaceous glands and fragmentation of hair shaft which is later replaced by fibrosis. Dermal heating also induces collagen remodeling. The combination of 1,450-nm diode laser with 1,550-nm erbium-doped fiber laser has potential for treating Nevus comedonicus.^{12,13}

CONCLUSION

Nevus comedonicus was presented for its rarity and for its unique presentation following Blaschko's lines.

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