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Case Report

Multi-dermatomal unilateral nevus comedonicus along the lines of Blaschko: a rare presentation

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ABSTRACT

Nevus comedonicus is a rare skin disorder due to defect in development of hair follicle presenting as groups of closely set, dilated follicular openings with dark keratin plugs resembling comedones. The lesions may be distributed in a linear, interrupted, unilateral or bilateral pattern and sometimes follow the lines of Blaschko. It may develop inflammatory acne-like lesions leading to cysts, abscesses and scarring. A 36 year-old woman presented with multiple pits on left half of the body involving the face, upper limb, shoulder, back and buttock since the age of 4 years. She also had a painful swelling over the back. There was no history of cranial, skeletal or ocular involvement. Physical examination revealed multiple circular honeycomb-type pitted scars sized from 0.1×0.1 to 0.5×0.5 mm present unilaterally along lines of Blaschko in an interrupted pattern over the left half of the body as mentioned above. Among these pitted scars few scattered comedones were present with interfollicular normal skin. There was an abscess along the margin of one of the lesions over the back which was tender, indurated and with local rise of temperature. Histopathological examination revealed deep, wide invagination of acanthotic epidermis, filled with concentric lamellae of keratin. Hair shafts were occasionally seen in the dermis, below the lower part of invagination. Based on clinical and histopathological picture a diagnosis of nevus comedonicus was made. She was treated with topical tretinoin and oral retinoids without much improvement. Nevus comedonicus in itself is a rare epidermal nevus, its multi-dermatomal unilateral distribution along the lines of Blaschko is even rarer.

Keywords: Nevus comedonicus, Lines of Blaschko, Multi-dermatomal

INTRODUCTION

Nevus comedonicus was first described in 1895 by Kofmann and termed it as "comedo nevus."[1] It is also called as follicular keratotic nevus, since there are no true comedones. It is caused by a defect in development of the hair follicle along with genetic mosaicism manifesting as groups of closely set, dilated follicular openings with dark keratin plugs resembling classical comedones. The plugged ostia have lamellated keratinaceous material which is not possible to be removed mechanically. Its pathogenesis may be due to over stimulation of FGFR2 signaling with increased expression of interleukin-1α.[2] It commonly affects face and neck area. Lesions may develop any time from birth to middle age, but are usually present at birth or develop before the age of 10 years. They may be distributed in a linear, interrupted, unilateral or bilateral pattern and sometimes follow the lines of Blaschko.[3,4] There is no sex or racial predilection. It may develop inflammatory acne-like lesions which can lead to cysts, recurrent bacterial infections, abscesses and scarring.[5,6] It may be part of nevus comedonicus syndrome in association with skeletal or central nervous system anomalies, ocular abnormalities, and cutaneous defects.[7] It may manifest with other skin disorders like ichthyosis, lichen striatus,

linear morphea, Sturge-Weber syndrome or hemangiomas, limb deformities like absence of fifth finger, polysyndactyly or clinodactyly. Central nervous system abnormalities may present with convulsions, dysgenesis of corpus callosum, electroencephalographic abnormalities or even paresis.[8] Nevus comedonicus syndrome has been observed to be associated with few epithelial tumors like pilar sheath tumour, syringocystadenoma papilliferum, keratoacanthoma and even sometimes tricho-epithelioma.[9,10]

CASE REPORT

A 36 year-old woman presented with multiple pits on left half of the body involving the face, upper limb, shoulder, back and buttock from the age of 4 years. She also had a painful swelling over the back. There was no history suggestive of cranial, skeletal or ocular involvement. There was no history of similar skin lesions in the family. Physical examination revealed multiple circular honeycomb-type pitted scars ranging in size from 0.1×0.1 to 0.5×0.5 mm present unilaterally over the left half of the body as mentioned above in interrupted linear pattern along the lines of Blaschko (Figure 1 to 3). Among these pitted scars few scattered comedones were present. The interfollicular skin was normal. There was an abscess over the back which was tender, indurated and with local rise of temperature (Figure 4). Nails and teeth were normal. There were no skeletal deformities. Histopathological examination revealed deep, wide invagination of acanthotic epidermis, filled with concentric lamellae of keratin. Hair shafts were occasionally seen in the dermis, below the lower part of the invagination (Figure 5). Based on clinical and histopathological picture a diagnosis of nevus comedonicus was made. She was treated with topical tretinoin and oral retinoids without much improvement.

Figure 1: Multiple circular honeycomb-type pitted scars ranging in size from 0.1×0.1 to 0.5×0.5 mm^3 present unilaterally over the left half of back along the lines of Blaschko. An abscess is also present over outer border of the lesions.

Figure 2: Multiple pitted scars over left half of face, neck and left upper limb.

Figure 3: Lateral view showing multiple pitted scars over left of the body with an abscess over the back.

Figure 4: Close up view of pitted scars with an abscess.

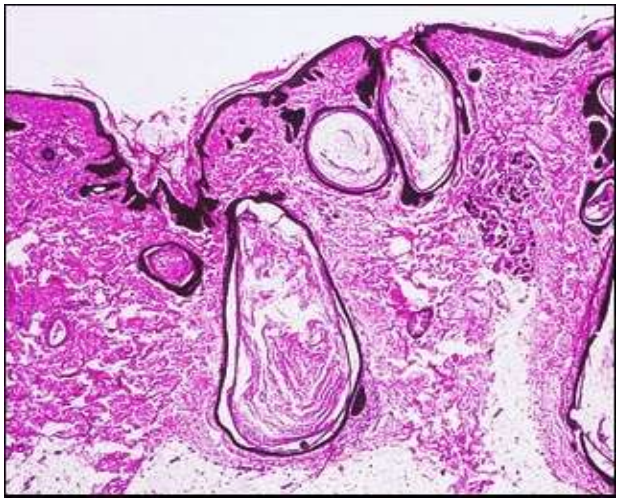

Figure 5: Histopathology shows deep and wide invagination of acanthotic epidermis which is filled with concentric lamellae of keratin.

DISCUSSION

Our patient's case is of interest in that it showed unilateral disseminated comedo-like lesions with complications of abscess formation. The skin lesions were clinically and histopathologically compatible with nevus comedonicus. We considered other diseases showing multiple comedo-like lesions such as disseminated congenital comedones, idiopathic disseminated comedones, chloroacne and multiple congenital comedones. The histopathological differentials were trichofolliculoma, dilated pore of Winer and folliculocystic and collagen comedo hamartoma. Disseminated congenital comedones are usually seen very early in life as they were reported by Galvan in a 3-month-old boy and confined only to the upper half of the body.[11] The distribution and age of onset of lesions differentiates our case from Disseminated congenital comedones. Idiopathic disseminated comedones is a very close differential for our case. It presents with symmetrically scattered comedo-like hyperkeratotic papules on the trunk, arms and legs sparing the face and neck which are the most common sites for Nevus Comedonicus.[12] On histopathology, crateriform cysts containing hyperkeratotic debris and peri-infundibular inflammatory infiltrate are seen which were not evident and seen in our histology specimen. Next differential for our case is chloracne which is caused by halogenated chemical compounds, it presents with diffuse acneiform lesions with diffuse grayish skin pigmentation and is a well-known entity after exposure of halogenated compounds but unfortunately our patient did not have any history of exposure to such compounds or any other acnegenic substances.[13] Multiple congenital comedones is a condition which presents at birth with congenital bilateral comedones associated with systemic complications.[14] Trichofolliculoma and dilated pore of winer were differentiated histopathologically with presence of only open 'Mother' follicles and absence of cysts from nevus comedonicus which has numerous open follicles as well as cysts.[15,16] Abundant collagen formation has been observed in folliculocystic and collagen comedo hamartoma on histopathology and also lesions are absent over face and neck unlike nevus comedonicus. After excluding the above mentioned disorders, we concluded that 'multi-dermatomal unilateral nevus comedonicus along the lines of Blaschko' is the most appropriate term to describe our patients condition.

CONCLUSION

Nevus comedonicus is a rare epidermal nevus. The hyperkeratotic epidermis with acanthosis makes the comedones extraction difficult compared to acne comedones. Diagnosis was based on clinical examination and histopathology. We are presenting a rare case of multi-dermatomal unilateral nevus comedonicus along the lines of Blaschko which is even rarer.

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