Zosteriform Nevus Comedonicus

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INTRODUCTION

Nevus comedonicus is a not so common benign hamartoma characterized by a closely set band of slightly elevated papules topped with darkened keratin plugs arranged in linear or zosteriform groups giving a sieve like appearance. It is due to a defect in the embryologic development of the hair follicle, aplasia or hypoplasia of the sebaceous gland (Carneiro et al., 1972; Sweitzer and Winer, 1932). These lesions appear mostly at birth or childhood (Givan et al., 2010; Guldbakke et al., 2007). Incidence of nevus comedonicus is equal in both males and females. Many factors like Gamma secretion, filaggrin, NEK somatic mutation, abnormal FGF2 signaling were recognized to contribute to the development of nevus comedonicus. Distribution of lesions is usually unilateral with a honeycomb like appearance of keratotic plugs. Benign tumors may develop over the pre-existing lesion. A detailed systemic examination mainly ophthalmic, skeletal and nervous system is mandatory in patients with nevus comedonicus to rule out syndromic manifestations. Dermascopy and histopathology aids in the diagnosis. There is no specific treatment for nevus comedonicus. Treatment is often unsatisfactory.
Till date, few cases of nevus comedonicus has been reported. Spontaneous resolution of lesions has not been reported in the literature. In our case, lesions are present unilaterally involving L2L3 dermatome with zosteriform distribution since the age of 6 years with no extracutaneous associations.

**Case Report**

A 30-year old female patient presented with complaints of multiple raised skin lesions with pits on her right lower limb since childhood. Initially, the patient developed a lesion on her right gluteal region at the age of 6 years. Gradually it progressed to involve the medial aspect of right thigh forming depressions filled with black thickened plugs. It was not associated with pain, itching, redness, discharge.

The patient was treated with multiple topical agents, but none of them was effective. No history of similar lesions elsewhere in the body. No history of similar lesions in the family. Dermatological examination revealed multiple, 1-3 mm greyish black clustered, discrete pits and dilated hair follicles filled with hyperkeratotic plug on the medial aspect of the right thigh (Figure 1) extending laterally in a linear pattern to involve the right buttock region giving the characteristic sieve-like appearance involving L2L3 dermatome (Figure 2).

**RESULTS AND DISCUSSION**

Nevus comedonicus, first described by Kofmann, is a rare variant of epidermal adnexal hamartoma which arises due to a developmental defect of mesodermal component of pilosebaceous unit as a result of which there is the inability to make properly formed hair matrix cells or sebaceous glands (Beerman and Homan, 1959; Kofmann, 1895). Incidence is equal in males and females. The prevalence has been found to be between 1 in 45,000 to 1 in 100,000. Genetic mosaicism plays a role in the etiology of nevus comedonicus (Liu et al., 2018; Levinsohn et al., 2016). Individual lesions are multiple aggregated papules with open comedo like lesions topped with black colored keratin plugs. Lesions are mainly located on the face, neck, upper arms, chest and abdomen. Other rare sites include the scalp, palms, soles, female genital area and glans penis (Wood and Thew, 1968; Abdel-Aal and Aziz, 1975).

![Figure 1: Multiple greyish grouped pits with few areas of hypo pigmentation giving a sieve like appearance.](image1)

Few skin colored nodules capped with comedones were scattered among these lesions. On palpation, lesions were non-tender and firm inconsistency. Scalp, oral mucosa, palms, soles, nails and genitalia were normal. Systemic examination was found to be normal. Clinical diagnosis of nevus comedonicus was made and skin biopsy was done to confirm the diagnosis. Section showed focal epidermal thinning, increased pigmentation in the basal layer and underlying dermis showed infundibular follicular plugging with rudimentary hair follicle (Figure 3).

![Figure 2: Band of dilated hair follicles plugged with keratin material distributed in zosteriform pattern involving L2L3 dermatome.](image2)
Neurological abnormalities include seizure disorders, mental retardation, microcephaly, transverse myelitis, corpus callosum dysgenesis, Sturge weber syndrome. Skeletal abnormalities comprise spina bifida, scoliosis, syndactyly, clinodactyly, rudimentary toe, (Alpsoy et al., 2005) bone cyst, bone hyper trophy, vit D resistant rickets (Seo et al., 2008).

Figure 3: Section shows epidermal thinning, increased basal layer pigmentation and rudimentary hair follicle.

The most common ocular change is cataract and oligodontia is a commonly reported dental abnormality. Nevus comedonicus syndrome is associated with other cutaneous disorders like ichthyosis, leukoderma, lichen striatus, linear morphea, hemangioma and hidradenitis suppurativa. Certain epithelial tumors like trichoepithelioma, keratoacanthoma, syringocystadenoma papiilliferum have found to be associated with NCS. Dermascopy is a useful tool for rapid diagnosis demonstrating numerous light and dark brown, circular or barrel-shaped, homogeneous areas with prominent keratin plugs (Vora et al., 2017). On histopathologic examination, each comedo is represented by a wide, deep invagination of the epidermis with keratin. These invaginations resemble dilated hair follicles. They actually represent rudimentary hair follicles. Occasionally one or more hair shafts are found in the lower portion of an invagination. One or two small sebaceous gland lobules may be seen in the lower pole of invagination. The keratinocytes show typical changes of epidermolytic hyperkeratosis. Diagnosis is usually clinical based on history (early age of onset) and typical morphology. Differential diagnosis includes comedonal acne, chloracne, acne nevus, familial dyskeratotic comedones, porokeratotic eccrine ostial dermal ductal nevus (PEODDN), scarring followed by herpes zoster, linear basal cell nevus, basal cell nevus with comedones, atrophoderma vermiculata, keratosis pilaris atrophicans, zosteriform darier’s disease, Favre-Racouchot Syndrome (nodular elastosis with cysts and comedones). There is no specific treatment for nevus comedonicus. Treatment is required for nevus comedonicus associated with inflammatory changes and for cosmetic purposes. Various treatment options are topical keratolytics like salicylic acid, retinoids, calcipotriene and ammonium lactate, dermabrasion, manual comedo extraction, surgical excision (Loria and Hailey, 1961). Small lesions can be completely excised. Successful treatment with CO2 lasers has been documented. In case of extensive nevus comedonicus, tissue expanders can be used. Topical management has a varying degree of improvement with a high recurrence rate.

CONCLUSIONS

Nevus comedonicus is a rare cutaneous developmental defect affecting the face, arms and trunk, which are the typical sites of occurrence. A deeper understanding of pathogenesis may lead to the evolution of many targeted therapies. Patients should be enlightened regarding the treatment options available and counseled about the lack of evidence predicting success with one treatment modality over the other. In our case, nevus comedonicus (type -1, munro’s acne nevus) is located on the thigh which is not a usual site of this condition and there were no associated systemic manifestations. The patient was suggested CO2 laser initially for debulking followed by surgical excision.

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Conflict of Interest

The authors declare that they have no conflict of interest for this study.

REFERENCES

Thillaikkarasi A et al., Int. J. Res. Pharm. Sci., 2020, 11 (SPL4), 2609-2612


