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Case Report

A rare epidermal naevus: naevus comedonicus case repot

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ABSTRACT

Naevus comedonicus (NC) is a rare epidermal naevus characterized by a honeycomb-like dilated follicular opening filled with black or brown keratinous plug. Diagnosis is based mainly on clinical examination and histopathology. It is also a part of NC syndrome (a neurocutaneous syndrome) with ophthalmological (congenital cataract), skeletal (scoliosis, fused vertebra, spina bifida, absence of the fifth finger, polysyndactyly, clinodactyly, rudimentary toe, bone hypertrophy, bone cyst, kyphosis and vitamin D resistant rickets), neurological (mental retardation, corpus callosum dysgenesis, seizures, paresis, electrocochleographic abnormalities). The physician should be aware of the extracutaneous manifestations involving the skeletal system, ophthalmological, neurological, and other skin manifestations. Several topical creams and energy-based devices treat with an unsatisfactory or moderate response. Surgical excision offers good aesthetic results.

Keywords: NC, Follicular naevus, NC syndrome, Adnexal skin tumours, Comedo nevus, Hamartoma

INTRODUCTION

Naevus comedonicus (NC), also known as Follicular naevus, is a rare epidermal naevus characterized clinically by honeycomb-like dilated follicular opening filled with black or brown keratinous plug. It is also a part of NC syndrome (a neurocutaneous syndrome) with ophthalmological, skeletal, spinal and neurological abnormalities.[1] Neavus comedonicus syndrome (NCS) term was coined by Peter B. Engber in 1978. NC was first described in 1895 by Kofmann, who used the term "comedo nevus".[2] NC can be present at birth or develop later in life, most commonly at the age of 10 years. Men and women are equally affected, and there is no racial predilection. Several topical creams and energy-based devices treat unsatisfactory or moderate responses.

CASE REPORT

A 43-year-old Indian woman, a housewife, presented to our OPD with multiple skin-coloured palpable lesions with black dots in the right side of the trunk, which were present since childhood. The lesions were asymptomatic except for occasional pain, swelling and discharge over the lesions.

The current episode was for four days. The clinical aspect of the lesion caused psychosocial distress during childhood but is not much of concern presently except for the episodes of infection. She is a homemaker, studied till higher secondary school. On dermatological examination, there was multiple dilated honeycomb type pitted scar filled with black coloured plug following blaschkoid pattern over the right side of the trunk (Figure 1 and 2). There was no history of seizures, ophthalmological, skeletal abnormality on examination. Routine haematological investigation including complete blood picture, liver function test, renal function test, X-ray spine was done and were in normal limits. CT head was not done since there were no signs and symptoms of neurological involvement clinically.

Based on the history and examination, skin lesions were diagnosed as NC. Since the patient was not worried about

the cosmetic appearance of the naevus, it was not treated. A course of antibiotics was given for reducing the swelling and inflammation.

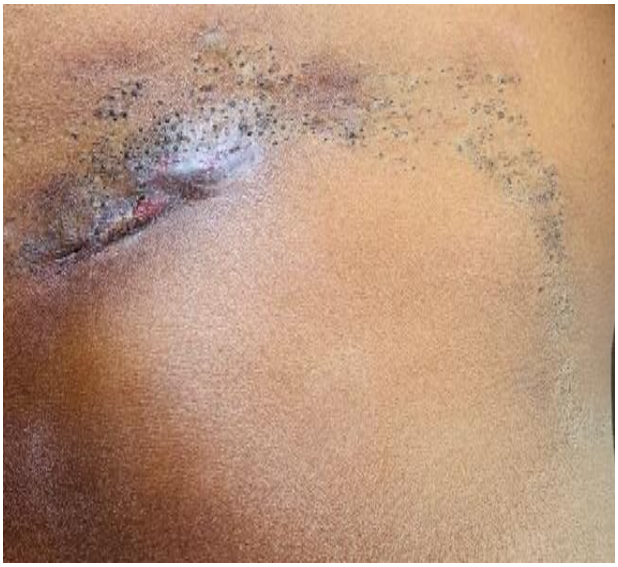

Figure 1: Multiple dilated honeycomb type pitted scar filled with black coloured plug following blaschkoid pattern over the right side of the trunk.

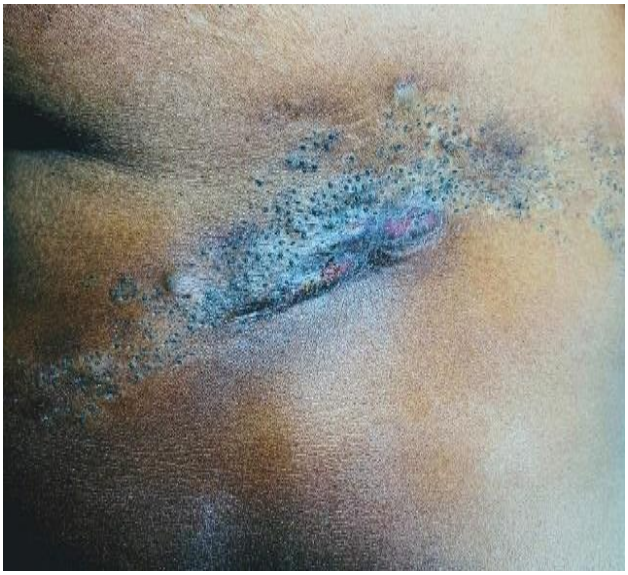

Figure 2: Multiple dilated honeycomb type pitted scar filled with black coloured plug with inflamed nodules with crusting.

DISCUSSION

NC is a rare naevus, first described by Kofmann in 1895, who used the term "comedo nevus".[2] NC clinically is characterized by multiple grouped dilated plugged Ostia filled with a keratinous plug. NC lesions present with various distribution patterns: linear, segmental, blaschkoid, unilateral, bilateral and extensive.[3] Most common affected sites are the face, neck, arm, chest, abdomen. Rare sites reported are the scalp, genitals, palms and soles.[4,5] NC skin lesions can occur at birth or develop later in life, most commonly developing before ten years. The estimated prevalence of NC is one in 45,000-1,00,000 population.[3] There is no gender or racial predilection. Clinically two types of NC are described in the literature: (1) non-pyogenic NC, which is characterized by comedones like plugs without suppuration, and (2) the second one is characterized by recurrent infections, cyst, pustules, fistulas and abscesses at various stages of development.[3,6]

NC can occur in combination with extracutaneous manifestations involving the skeletal system, ophthalmological, neurological, and other skin manifestations. This combination subtype is part of NCS, and the term was coined by Peter B. Engber. NCS belongs to the group of epidermal Naevus syndrome. Apple used the word "epidermal naevus syndrome" to include six disorders having epidermal naevus (verrucous epidermal naevus, sebaceous naevus, NC) with extracutaneous manifestation. Extracutaneous manifestations are ophthalmological (congenital cataract), skeletal (scoliosis, fused vertebra, spina bifida, absence of the fifth finger, polysyndactyly, clinodactyly, rudimentary toe, bone hypertrophy, bone cyst, kyphosis and vitamin D resistant rickets), neurological (mental retardation, corpus callosum dysgenesis, seizures, paresis, electrocochleographic abnormalities).[3,7] Other skin disorders Klippel Trenaunay, Sturge-Weber syndrome, trichoepithelioma, the dilated pore of Weiner, Linear morphea, lichen striatus, keratoacanthoma, syringocystadenoma papilliferum and rarely squamous cell carcinoma and basal cell carcinoma.[3,8-11]

The exact pathophysiology of NC is not known. Mutation in the fibroblast growth factor receptor 2 (FGFR2) is considered to be an essential factor in the pathogenesis of NC.[3] FGRF2 belongs to the family of tyrosine kinase receptors. FGFR2 overstimulation results in increased expression of interleukin-1-alpha, which may be involved in the pathogenesis. Recent studies have highlighted the mutation in NEK 9 and absence of gamma-Secretase in NC.[12,13]

On histopathological examination, each comedon is represented by a wide, deep, dilated invagination of epidermis filled with keratin and devoid of the hair shaft. This invagination is thought to be a rudimentary hair shaft. Occasionally one or two small sebaceous glands can be seen opening into the invagination. Epidermolytic hyperkeratosis can be seen in the epidermis.

Several topical creams and energy-based devices are used for the treatment with an unsatisfactory or moderate response. NC is a benign skin lesion and does not require aggressive treatment, and as with other epidermal naevi, its treatment is quite challenging. NC should be treated not only for cosmetic purposes but also for the infection and scars.

Topical creams like emollients, moisturizer, topical retinoids (tazarotene), 12% ammonium lactate, salicylic acid, topical corticosteroids (for inflammatory lesions), topical tacalcitol with calcipotriene, have been tried with a reasonable response.[14-16] Oral isotretinoin is found to be effective in widespread inflammatory acneiform naevi.[17] Localised NC can be removed by surgical excision with an excellent aesthetic result if the size and site of the lesion permits. Comedon extra traction, superficial shaving, dermabrasion have been tried. Laser treatments with erbium YAG and ultra-pulse CO_2 have shown cosmetic response in single patients.

CONCLUSION

NC is a rare epidermal naevus characterized by honeycomb-like dilated follicular opening filled with black or brown keratinous plug. Diagnosis is based mainly on clinical examination and histopathology. The physician should be aware of the extracutaneous manifestations involving the skeletal system, ophthalmological, neurological, and other skin manifestations. Several topical creams and energy-based devices are used for the treatment with an unsatisfactory or moderate response.

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