

Egyptian Dermatology Online Journal

Volume 5 Number 2

Nevus comedonicus in a Malay man: A case report and review of literature

Felix Boon Bin Yap, MD (UNIMAS) MRCP (UK); Pubalan Muniandy, MBBS (Malaya) MRCP (UK)

Egyptian Dermatology Online Journal 5 (2): 12

Department of Dermatology, Sarawak General Hospital, Jalan Hospital, 93586
Kuching, Sarawak, Malaysia

e-mail: woodzlamp@yahoo.com

Submitted: October 13, 2009.

Accepted: November 19, 2009.

Abstract

Nevus comedonicus is a rare adnexal hamartoma of the pilosebaceous units. Here, a 25 year old Malay fireman with nevus comedonicus is presented. His nevus comedonicus, comprising of a linear group of hyperpigmented comedo like papules is seen on the right side of his neck. There was no associated pain, itching or discharge. He presented mainly for cosmetic reason. Skin biopsy of the lesion showed the presence of widely dilated invagination resembling dilated hair follicle filled with orthokeratotic horny materials penetrating the reticular dermis. The interfollicular epidermis was normal. Treatment with keratolytics and topical tretinoin was not satisfactory. He had successful excision of the lesions.

Introduction

Nevus comedonicus is a rare adnexal hamartoma involving the pilosebaceous units. It was first reported in 1895 by Kofmann [1]. Clinically, it presents with linear groups of asymptomatic open comedo-like pits filled with black keratinous plugs [2,3]. Both unilateral and bilateral distributions are seen [2,4,5]. Face is the most commonly affected site followed by the neck, the trunk and the upper arms [2].

Here, a case of nevus comedonicus involving the right side of the neck in a 25 year old Malay man from Miri, Sarawak is reported.

Case report

A 25 year old man presented with multiple papular lesions on the right side of the neck since childhood. He presented to the skin clinic because he was embarrassed of the lesions. He was always teased by fellow colleagues in the fire and rescue

department where he works. The lesions were not painful or itchy. There was no discharge. No other family members had similar skin lesions.

On examination, there were linear, grouped, hyperpigmented, comedo like, keratotic papules on the right side of his neck in an area measuring 4 cm X 3 cm (**Figure 1**). There were no other lesions on his body



Fig 1: Linear group of comedo-like hyperpigmented papules on the right side of the neck.

Skin biopsy of one of the comedo like papule showed deep and wide invagination resembling dilated hair follicle lined by epithelium and filled with orthokeratotic horny materials penetrating the reticular dermis (**Figure 2, 3**). The interfollicular epidermis was normal. This finding was consistent with the diagnosis of nevus sebaceous.

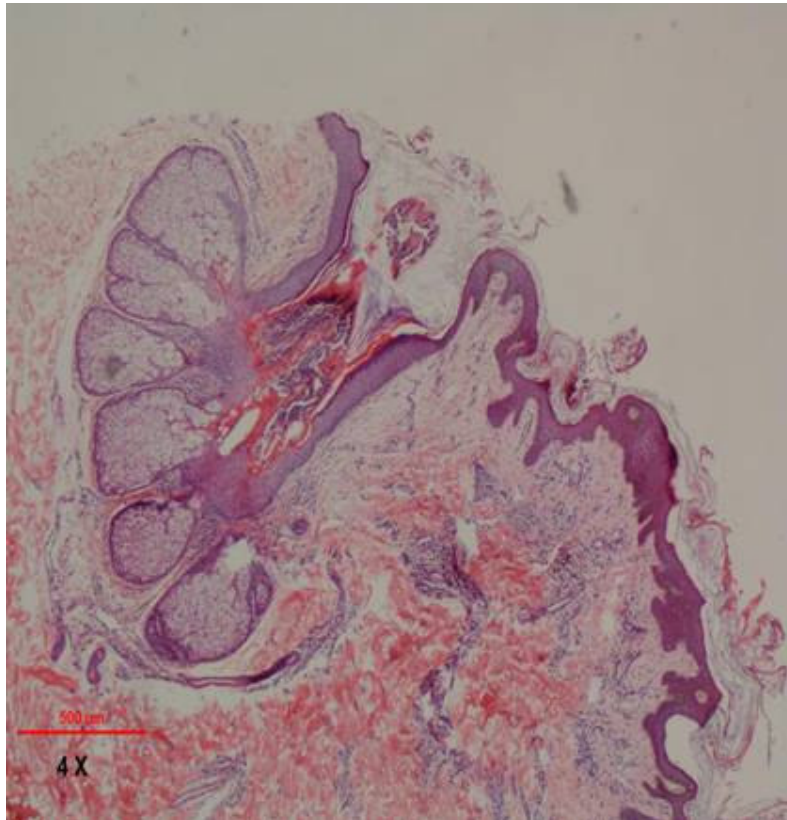


Fig 2: Presence of widely dilated invagination resembling dilated hair follicle filled with orthokeratotic keratinous plug with perifollicular hyperkeratosis, acanthosis and papillomatosis. Hematoxylin and eosin. Magnification X 4.

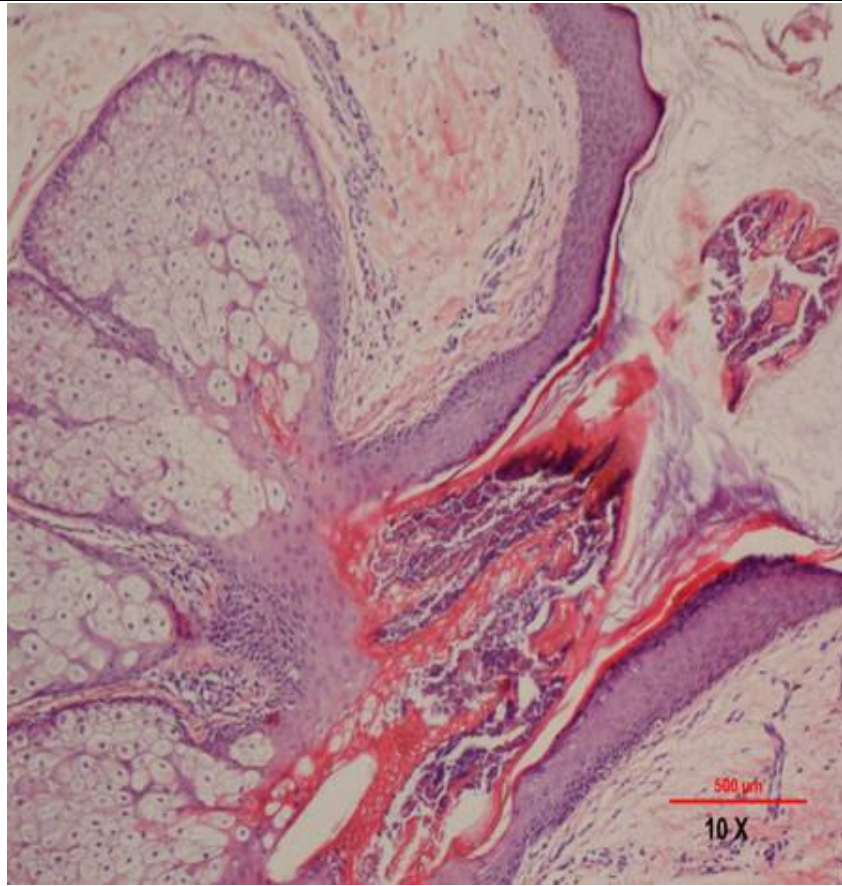


Fig 3: Close up view of the orthokeratosis.

He was given topical 20% salicylic acid ointment and topical tretinoin locally for the lesions. On follow up 2 months later, no satisfactory regression was seen. He had successful excision of the lesions.

Discussion

Nevus comedonicus is an uncommon developmental anomaly also known as nevus acneiformis, zoniform nevus with comedones and systematized sebaceous gland nevus [4]. It is reported in all racial groups. It affects both sexes equally. It could be present at birth but is usually seen in childhood. Most patients present before the age of 10 [6]. Familial cases of nevus comedonicus are seen with variable mode of inheritance [7].

The etiology of nevus comedonicus is unknown. Some consider it as a hamartoma arising from a defective mesoderm, while others consider it to be an epidermal nevus involving the appendages namely the hair follicle and the sweat ducts [8,9,10]. Proponents of defective mesodermal theory postulated that there is developmental failure of the mesodermal pilosebaceous complex resulting in imperfect differentiation of the epidermal component. The follicular structure that forms is unable to generate hair matrix cells. Nevertheless, it is able to produce the soft keratin that plugs the adnexal orifice, forming the comedones [2,11]. Those

favoring the epidermal nevus theory found epidermolytic hyperkeratosis, acanthosis and papillomatosis in histopathology, findings that are characteristically seen in epidermal nevus [12].

Clinically, nevus comedonicus can present in 2 clinical entities. The first is the asymptomatic comedo like eruptions and the second is the presence of cysts, scars, fistulas, and abscesses due to inflammation and infection of the comedo like lesions [3,13]. The comedo like eruptions consist of asymptomatic, grouped, usually 20 to 50 small, pin head, hyperpigmented papular lesions. Small comedones appear as keratin filled pits while larger ones extend above the surface of surrounding skin and give a 'nutmeg crater' feeling. The keratin material can be shelled out freely, leaving craters in the normal skin. These lesions are usually seen on the face, neck, trunk and upper arm. Albeit rare, there are also reported on the palms, scalp, ears and genitalia [14,15,16,17]. The lesions can cover an area as small as 2 cm to extensive involvement of the body [2,4,5,18,19]. Most of the lesions are unilateral and segmental, following the lines of Blascko [4].

Approximately 50% of patients with nevus comedonicus will present with the second clinical entity. The recurrent inflammation and infection in this entity poses a more severe clinical problem for both the patients and the treating physician as it will lead to abscesses, cysts, fistulas and eventually scars; as opposed to the mainly cosmetic problem poses by the first entity. This entity is more commonly seen in adolescents and adults as the inflammatory disease activity and severity increases with age [3].

Follicular tumors have been reported to occur within the nevus comedonicus. These tumors include trichofolliculoma and pilar sheath acanthoma [20,21]. Epidermoid cyst, trichilemmal cysts, linear cell nevus, ichthyosis and Alagille syndrome have also been associated with this condition [22,23,24,25,26]. Association with extracutaneous manifestations especially neurological, ocular and skeletal anomalies have been termed nevus comedonicus syndrome [5,27]. These anomalies include electroencephalographic abnormalities, ipsilateral cataract, corneal changes, hemivertebrae, scoliosis and absence of the fifth ray of a hand [28].

Histopathological examination shows large dilated pilosebaceous follicles filled with orthokeratotic keratin materials [4,13]. One to several hair shafts can be found occasionally on the base of the lesion. Similarly, small sebaceous gland lobules may be seen opening into the lower pole of the invaginations [4]. The interfollicular epidermis shows hyperkeratosis and papillomatosis resembling epidermal nevus. Occasionally, epidermolytic hyperkeratosis is seen [12]. Inflamed and infected lesions will show presence of dermal inflammatory cells infiltrates.

Nevus comedonicus needs to be differentiated from linear epidermal nevus, comedonal acne, lymphangioma and lichen striatus clinically. The differential diagnosis for the inflamed entity includes inflammatory linear verrucous epidermal nevus and its variants; linear lichen planus and linear psoriasis. Differentiation of nevus comedonicus with other epidermal nevi is easy as the former shows presence of comedones, which on extraction will leave a crater on the skin surface. Finding groups of lesions following the lines of Blascko excludes comedonal acne. Lymphangioma circumscriptum usually presents as groups of vesicle like papules

resembling frog spawns, unlike the comedonal lesions of nevus comedonicus. Histologically, differentiation from comedonal acne is important. In comedonal acne, the pilosebaceous units are complete whereas those in nevus comedonicus are poorly formed. Moreover, in nevus comedonicus, hyperkeratosis and papillomatosis are frequently observed in the interpapillary epidermis and absent in comedonal acne. Dilated pore of Winer can sometimes be confused with nevus comedonicus histologically. However, this condition is usually seen in the elderly and can be differentiated clinically.

Treatment of nevus comedonicus is mainly for cosmetic reason. However, for those with the inflamed entity, proper treatment is needed to prevent complications of the recurrent infection and inflammation. Various types of treatment have been used to treat nevus comedonicus. Treatment outcomes are mostly unsatisfactory [4,15]. Topical agents like tretinoin, ammonium lactate lotion, tazarotene and calcipotriol have been reported to be cosmetically successful by some [29,30,31]. Pore strips to eliminate keratin plugs have also produced excellent cosmetic outcome in a Japanese patients [13]. Erbium-YAG laser have also been reported to be successful [32]. Dermabrasion, carbon dioxide lasers and comedo extraction have all been used with variable success [33]. Topical and systemic antibiotics are usually needed in the inflamed entity. Use of oral isotretinoin may partially suppress the inflammation but many fail to respond [34]. The comedonal lesions do not respond to oral isotretinoin. Surgical excision is useful especially in limited areas of inflammation [34].

In conclusion, nevus comedonicus is a rare developmental anomaly causing mainly cosmetic disfigurement. Complications due to recurrent inflammation and infection can occur. Treatment outcome both cosmetically and medically are variable. A classical case of nevus comedonicus with poor topical treatment outcome but successful surgical excision was presented.

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