

# Systematized Nevus Comedonicus Associated with Multiple Congenital Anomalies (Nevus Comedonicus Syndrome)

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## Introduction:

Nevus comedonicus represent developmentally abnormal and ectopic pilosebaceous follicles. It is not considered a hereditary disorder. However a diffuse familial form been reported. Usually appear at birth or during childhood. Nevus comedonicus comprise groups of pits filled with black keratinous plugs resembling blackheads, present with a circumscribed area. Areas of more marked background hypopigment patches following the Blaschko's lines have been reported by several authors. They may be or several lesions in a linear, unilateral or more rarely bilateral distribution. Extensive, systematized lesions were reported to be extremely rare. Common sites of involvement are the face, neck and trunk. Lesions on the palms, soles and penis has been reported. Associated Cutaneous finding with nevus comedonicus including: Inflamed pustules, Abscess, Epidermal cysts, Trichilemmal cysts, Background of hypopigmented or hyperpigmented patches, verrucous nodules, and scarring. Associated anomalies with nevus comedonicus have been reported including: Congenital cataracts, skeletal abnormalities (scoliosis, hemivertebrae, absence of the fifth digit), Ectodermal dysplasia, Extensive nevus flammeus, EEG abnormalities, Perforating elastoma. Nevus comedonicus syndrome is characterized by a nevus comedonicus in combination with ipsilateral cataract and skeletal defects. Electroencephalographic abnormalities have likewise been reported.

## Case Report

17-years-old, Saudi male patient was admitted to

A.C.H. under the derma case for investigation with 10-years history of asymptomatic, persisting, extensive, blackish skin lesions, involving face, neck, trunk, groin, buttocks and extremities.

History of persisting hypopigmented patches, scattered all over the body since birth. History of recurrent attacks of inflamed pustules and abscesses at the site of these skin lesions for the last few years. History of below average performance at school. No similar family history. There is history of consanguinity. Physical examination, patient looks mentally retarded. Vital signs were stable. No cyanosis, jaundice or lymphadenopathy. Chest, CVS, CNS and abdomen were normal. There is Asymmetry of the face and clavicles, Scoliosis, Pectus excavatum, V-shaped arched palate and multiple exostosis.

Cutaneous Examination showed extensive, scattered groups of pits filled with black keratinous plugs, resembling comedones, present with a circumscribed area. Most of these skin lesions appear on hypopigmented patch, involving face, neck, trunk, groin, buttocks and extremities (Figs. 1, 2, 3,4,5). Patient also has multiple, soft, non-scaly, skin color, nodular skin lesions, within a circumscribed area containing comedones, present on the ears, neck, groin and buttocks (Figs. 2,4). Scarring appears mainly in the groin lower abdomen and upper part of both thighs. These hypopigmented patches were following the lines of blaschko: Linear on extremities (Fig.5), S-shapes on abdomen (Fig.4), V-shaped on the posterior mid-line. Buccal mucosa was spared. Nails and hair were normal.

Clinical Impression was systemized nevus comedonicus associated with: Mental retardation, Trichilemmal or Epidermal cysts, skeletal abnormalities (Asymmetry of the face & clavicles, scoliosis, pectus excavatum & multiple exostosis). Extensive hypopigmented patches following the lines of Blaschko, Scarring and V-shaped arched palate.

Investigation: routine laboratory investigations were within normal limits including: complete blood counts, liver function test, urea and creatine, electrolytes, chest x-ray, electro-cardio gram. Ultrasound of abdomen was normal. Ophthalmological examination showed no abnormalities. Skeletal survey showed on

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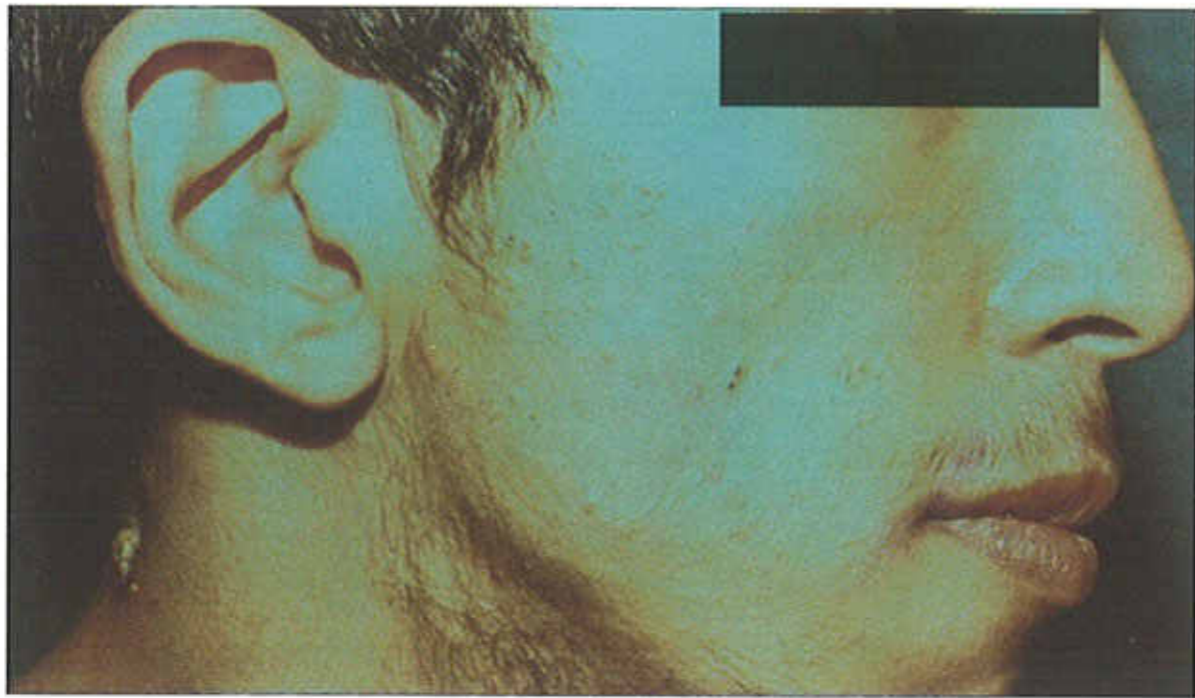


Fig. 1 Scattered groups of pits filled with black keratinous plugs, resembling comedones on the face.



Fig.2 Scattered groups of pits filled with black keratinous plugs, resembling comedones on the neck. Also, patient has multiple, soft, non-scaly, skin color, nodular skin lesions present on the ears and neck.



Fig.3 Scattered, extensive groups of pits filled with black keratinous plugs, resembling comedones, present with a circumscribed area. Most of these skin lesions appear on hypo-pigmented patch involving the trunk.



Fig.4 Comedones, presents on hypopigmented patches involving the trunk, groin and extremities following the lines of blaschko. Also, patient has multiple nodular skin lesions present in the groin and lower abdomen.



Fig.5 Comedones, present on hypopigmented patches were following the lines of blaschko. Linear on extremities.

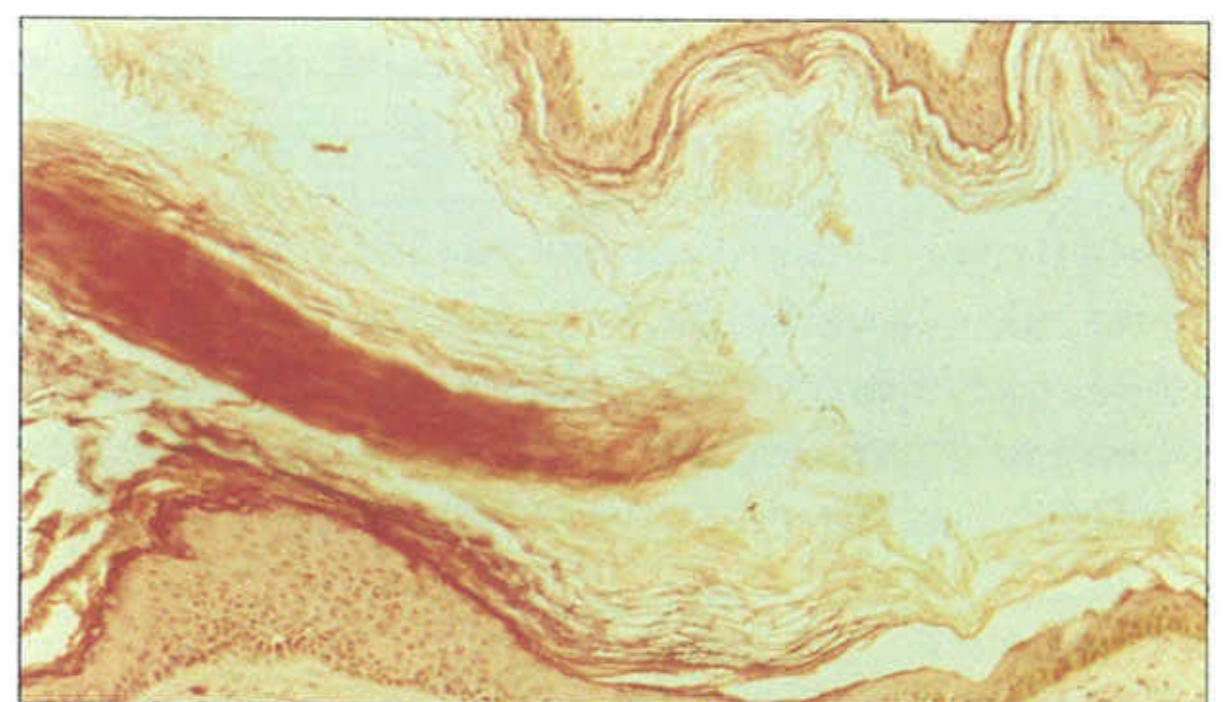
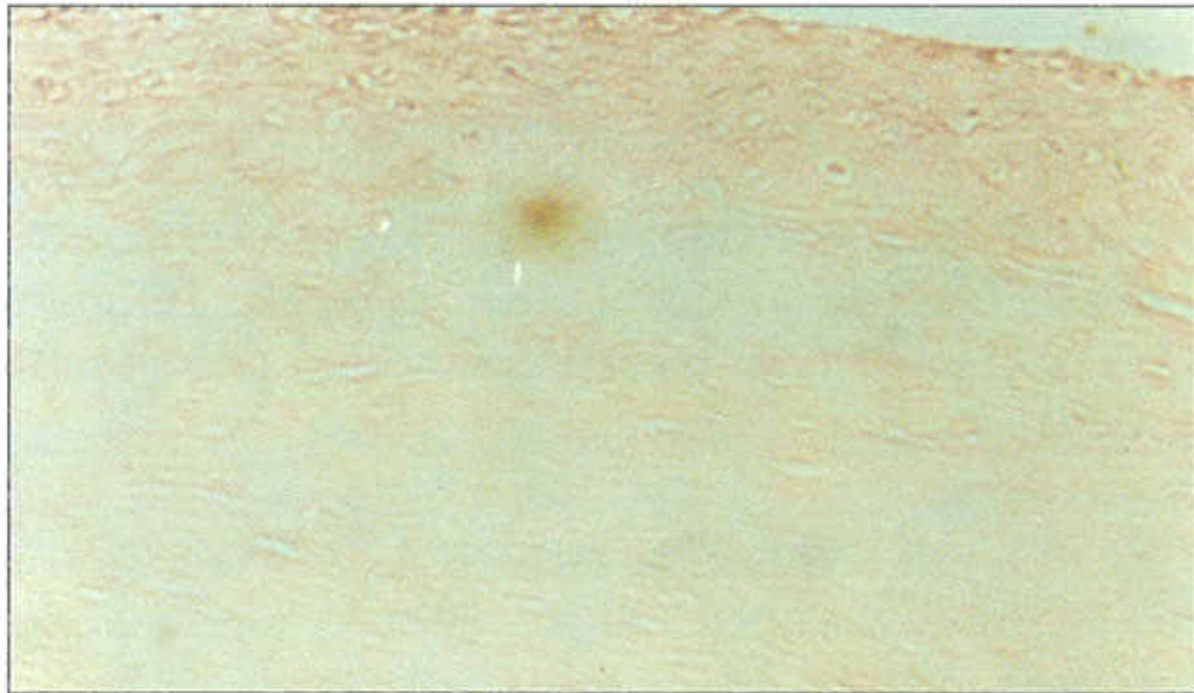


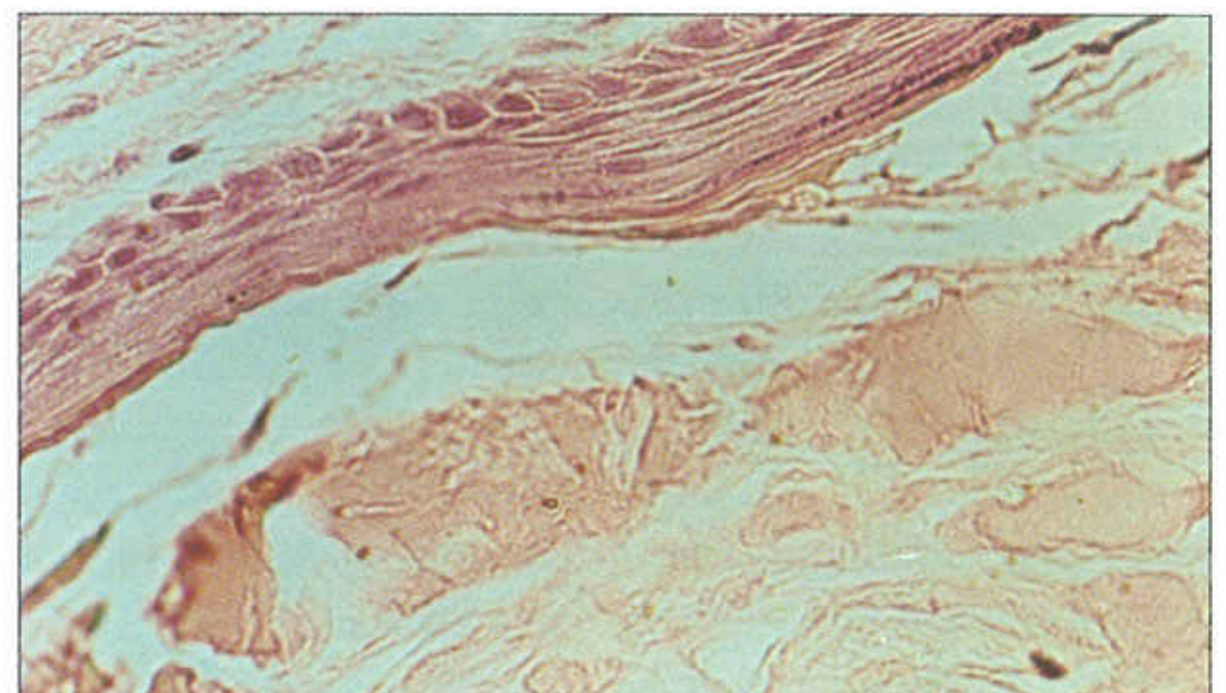
Fig.6 The histopathological results of skin biopsy from area contain the comedones showed deep and wide invagination of epidermis filled with concentric lamellae of keratin hair shaft can be seen in the lower part of the invagination.

x-ray findings: Scoliosis of the dorsal spine, Anterior wedging of lower dorsal and upper lumbar vertebrae, Peridiscal sclerosis, Multiple exostosis, CT Scan of the brain, showed Arachnoid cysts behind the thalamus,

Electroencephalographic (EEG) was normal. Skin biopsies were taken: From the right shoulder (area contains the comedones), Multiple nodular skin lesion were surgically excised from ears, neck and groin for



*Fig. 7 The histopathological results of multiple nodular skin lesions from neck and ears showed thick wall of stratified epithelium. The cell get larger toward the lumen and fade individually into the dense keratinized mass without formation of keratohyalin with trichilemmal cyst.*



*Fig. 8 The histopathological results of multiple nodular skin lesions from groin showed the cyst wall is composed of mature keratinized squamous epithelium. There is prominent granular cell layer and keratin lamelle in the lumen, consistent with epidermal cyst.*

histopathology. The histopathological results of skin biopsy from area contain the comedones showed a comedo with surface opening and fragmented shafts in its Lumen plugged with keratin consistent with nevus comedonicus (Fig.6). The histopathological results of multiple nodular skin lesions from neck and ears showed thick wall of stratified epithelium, the cell get larger toward the lumen and fade individually into the dense keratinized mass without formation of Keratohyalin, consistent with trichilemmal cyst (Fig.7).

The histopathological results of multiple nodular skin lesions from groin showed the cyst wall is composed of mature keratinized squamous epithelium. There is prominent granular cell layer and keratin lamellae in the lumen, consistent with epidermal cyst (Fig.8).

The final diagnosis was systematized nevus comedonicus associated with Mental retardation, Epidermal cyst, Trichilemmal cysts, Extensive background hypopigment patches, Scarring, Skeletal abnormalities (scoliosis, pectus excavatum, asymmetry of the face & clavicles and multiple exostosis, V-shaped arched palate and Arachnoid cysts behind the thalamus.

Treatment: No definite treatment, Topically, Tretinoin and 12% ammonium lactate were used. Also, regular use of comedo extractor and surgical excision were done for large trichilemmal and epidermal cyst.

#### **Discussion:**

The comedo nevus is a development abnormality of the skin comprising numerous keratin-filled pits. It has generally been assumed that comedo nevi are of pilosebaceous origin.

These nevi usually appear at birth, during childhood or adolescence at an onset as late as the seventh decade has been observed. (1,2) Frequently, their pattern of distribution is linear or band-like and skip areas within the nevus may be seen. Also unilateral (9,10,11) or more rarely, bilateral distribution has been reported (12). Lesions may be very extensive. (8,11) Common sites of involvement are the face, neck, trunk and upper arm but there have been reports of lesions on the palms as well as the penis. (3,4) Those cases with extensive involvement provide direct evidence that nevus comedonicus can follow Blaschko's Lines. (3,5,6) Associated Cutaneous findings within the nevus include abscesses, scarring, epidermal cysts, trichilemmal cysts, (13) alopecia, verrucous nodules, and background hypopigmentation or hyperpigmentation, (3,7,8) areas of more marked hypopigmentation have been reported by several authors. (3,8)

As with other types of epidermal nevus, there have been occasional reports of associated development anomalies, including ipsilateral cataract, (14) skeletal malformation (scoliosis, hemivertebrae, absence of the fifth digit), (8,15,16) extensive nevus flammeus, (17) transverse myelitis (15) perforating elastoma, (18) and ectodermal dysplasia. The most conspicuous histological feature of comedo nevi is the presence of deep and wide invaginations of epidermis filled with concentric lamellae of keratin. The invaginated epidermis is generally acanthotic. It is assumed that these generally represent dilated hair follicles, because hair shafts may occasionally be seen in the lower part of the invagination. (19,20,21)

Clinically, the differential diagnosis includes porokeratotic eccrine ostial and dermal duct nevi may closely resemble comedo nevi, and the two entities have

frequently been confused, particularly when the palm or sole is affected. Another entity, recently describe with a clinical appearance virtually identical to comedo nevus, but with distinctive histological features, is the dilated nevus.<sup>(22)</sup> Widespread comedones also occur symmetrical in the inherited disorder known as familial diffuse comedones.<sup>(23,24,25)</sup> Comedones may also occur in the rare type of epidermal nevus known as linear basal cell nevus. Atrophoderma vermiculata and keratosis pilaris atrophicans have been confused with comedo nevi, but are distinguished principally by their symmetry. Treatment may be sought either for cosmetic reasons or because of secondary inflammation. Asymptomatic lesion may be left untreated. As with other epidermal nevi, surgical excision will be more effective in the long term than superficial shaving or dermabrasion,<sup>(9,26)</sup> but the lat-

ter can occasionally be useful cosmetically if the lesions are extensive.<sup>(27)</sup> As can the regular use of as comedo extractor.<sup>(9)</sup> Both topical retinoic acid and 12% ammonium lactate may sometimes improve the appearance of lesions,<sup>(28,29)</sup> and may help prevent secondary inflammation. Oral isotretinoin has been reported to be ineffective.<sup>(30,31)</sup>

### Conclusion:

We present a very rare case of 17 years old, male Saudi patient with systematized nevus comedonicus associated with: Mental retardation (never reported before). Epidermal cysts, Trichilemmal cysts, Background of hypopigmented patches following the blaschko line, Scarring, Skeletal abnormalities (scoliosis, pectus excavatum, asymmetry of the face and clavicles and multiple exostosis), V-shaped arches palate. Arachnoid cysts behind he thalamus.

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