



International Journal of Homoeopathic Sciences

E-ISSN: 2616-4493

P-ISSN: 2616-4485

www.homoeopathicjournal.com

IJHS 2021; 5(2): 217-223

Received: 07-02-2021

Accepted: 09-03-2021

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Clinical description of a rare dermatological disease, acne Keloidalis Nuchae and successful homoeopathic management: A case report

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DOI: <https://doi.org/10.33545/26164485.2021.v5.i2d.385>

Abstract

Acne Keloidalis Nuchae (AKN), also known as Folliculitis Keloidalis Nuchae (FKN), is a rare dermatological disease characterised by persistent folliculitis at the nape of the neck that forms keloid like scars and finally leads to cicatricial alopecia, and mainly affects the African ethnic group ranging from 0.45 to 9% prevalence rate. The exact aetiology is indistinct, but it is supposed to be triggered by chronic irritation or occlusion of the hair follicles due to different causes, which leads to the inflammation of the affected parts. If untreated, fibrosis and keloid formation can be evident, presented with chronic scarring and/or scarring alopecia without active inflammation. Pruritis, pain, contact bleeding and alopecia are some of the clinical presentations of this illness. If the secondary bacterial infection is present, pustules, sinuses and abscesses may be seen. Conventional medical management includes topical or oral antibiotics, corticosteroids, retinoids, topical fusidic acid and topical urea, intralesional injection of triamcinolone, surgical excision, electrosurgery, cryotherapy, light and laser therapy. This article aims to deliver all the relevant information of AKN presented in literary works and present the successful case treated with *Calcarea Phosphoricum* and individualised Homoeopathic medicine. This article will highlight the need for the Individualisation of a case, although rare, required to treat any named ailment with the help of Homoeopathy.

Keywords: Acne Keloidalis nuchae (AKN), folliculitis Keloidalis nuchae (FKN), homoeopathy, *Calcarea phosphoricum*

Introduction

Acne Keloidalis Nuchae (AKN), also known as Folliculitis Keloidalis Nuchae (FKN), is a disease characterised by persistent folliculitis at the nape of the neck that forms keloid like scars and finally leads to cicatricial alopecia. The lesions arise as a result of folliculitis^[1]. In 1860, Hebra first used the terminology as a Sycosis Framboesiformis; in 1869, Kaposi mentioned a Dermatitis Papillary Capillitia, later Bazin in 1872 coined the term Acne Keloidalis Nuchae^[2].

Epidemiology

More in African ethnic groups, the prevalence of this illness varies between 0.45-9%, in Nigeria ranging from 0.7% to 9.4%, West Africa 0.7% and South Africa 4.7% in boys and 10.5% in older men. In African-Americans ranges from 0.5%-13.6%. In Asians, it is very low (0.007%). Moreover, the condition has a predilection for men, occurring 20 times more frequently than in women^[2-4].

Aetiology

The exact aetiology of AKN is unclear. However, it might be triggered by chronic irritation or occlusion of the follicles due to haircutting practices (e.g., close shaves), trauma, friction (e.g., rubbing from shirt collars or helmets), heat, or humidity as a predisposing or exacerbating factor. Also, it might be triggered by infection (Demodex or bacteria). Other potential contributory factors include autoimmunity, excess androgens or increased sensitivity to androgens, seborrhea, and medications (e.g., cyclosporine)^[5,6].

Pathogenesis

Due to repeated friction, trauma, and infection of the skin, overlying the nape of the neck leads to developing this illness. The natural course of disease starts with the early formation

of inflamed papules with marked erythema. Secondary infection can lead to pustules and abscess formation in some cases. Over time, continued inflammation leads to pronounced fibrosis and keloid formation with coalescence of the papules into large plaques and nodules. Later stages of presentation include chronic scarring and/or scarring alopecia without active inflammation [6].

Classification [7]

The proposed three-tier AKN classification was based on the lesion type and sagittal width (Figure1) based on lesion distribution, primary lesion type, and associated scalp disease. The distribution of area lies between two horizontal lines drawn at the level of the occipital prominence and the inferior tips of the mastoid processes. The occipital prominence was a prominent landmark because breaching it

removes the lesions from the generally concave zone that is naturally present in the nape-neck axis. The determination of class categories based on the clinical experience and results of the complications of the surgical dataset. The classification is as follows:

- Class, I lesions are confined to a 3-cm or less sagittal width within the area defined by the aforementioned clinical demarcation lines.
- Class II lesions are confined to a sagittal width of more than 3cm but less than 6.5cm, not breaching the demarcation lines.
- Class III lesions are greater than 6.5cm in size or breach clinical demarcation lines but are generally confined to the nuchal area.
- Class IV lesions are widespread and significantly exceed the nuchal area.

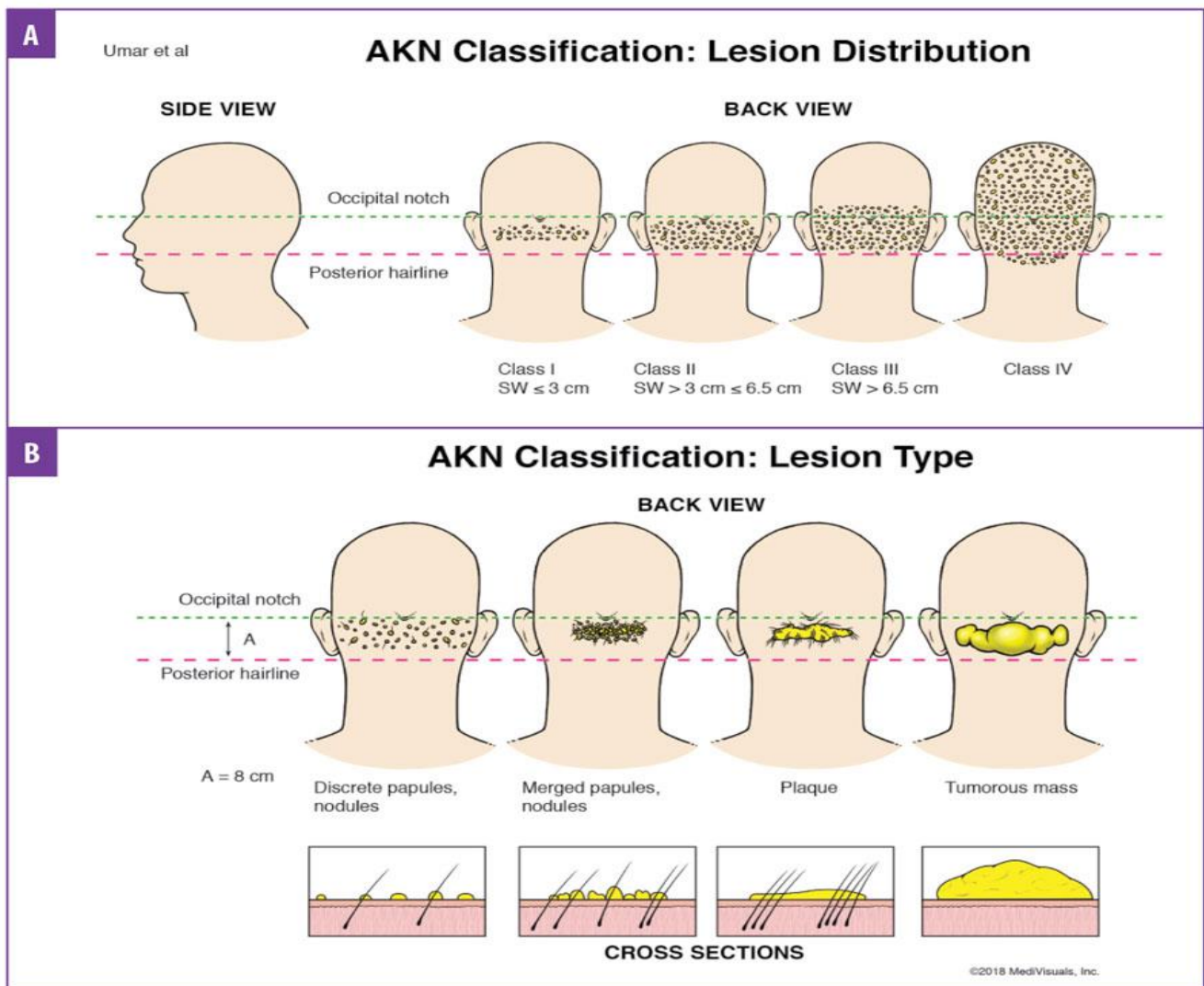


Fig 1: AKN classification Schema: A) AKN classification by lesion distribution of dominant lesions relative to an area in the occipital area that lies between two horizontal lines drawn at the level of occipital prominence and the inferior margins of the mastoid bone: B) AKN classification by primary morphology of the dominant lesions type relative to an area in the occipital area that lies between two horizontal lines drawn at the level of the occipital prominence and the inferior margin of the mastoid bone.*

[*Umar S, Lee DJ, Lullo JJ. A Retrospective Cohort Study and Clinical Classification System of Acne Keloidalis Nuchae. Journal of Clinical & Aesthetic Dermatology. 2021 April 1;14(4).]

Risk factors: [4, 8, 9, 10].

- Dark-type of skin and curly hair.

- Young age (commonly in adolescence/early adulthood).
- Male predominance.
- Hormonal influences (especially Testosterone).
- Family history.
- Excessive weight, obesity and diabetes mellitus.
- Frequent trauma to the scalp, e.g., wearing the helmet, Close shaving and haircutting procedures.

Clinical features [2, 11]

This illness develops primarily after puberty. Initially, varying sizes of the papules and/or pustules on the nuchal or/and occipital region extend laterally or sometimes towards the vertex.

The onset is usually preceded by pruritis a few hours to days after a haircut or after using headwear such as a helmet, followed by pain and contact bleeding. Pruritus sets up a cycle of itch, scratch, irritation, and inflammation. In addition, the secondary bacterial infection is common, predisposing to pustules, sinuses and abscesses, and if these lesions are more extensive, it affecting the patients' self-esteem.

In some individuals, it may be an initial generalised folliculitis healing with the residual lesion of AKN, which tends to recur. Recurrence of this folliculitis predispose to the destruction of hair follicles leads to patchy scarring alopecia. Kridin K Patel *et al.*, in their population-based study, demonstrated that this AKN has a significant association with Hidradenitis suppurativain an Israeli population.

Medical management [2, 12].

It includes topical or oral antibiotics, corticosteroids, retinoids, topical fusidic acid and topical urea, intralesional injection of triamcinolone, and surgical excision, electrosurgery, cryotherapy, light and laser therapy.

Prevention [6]

- Avoid frequent close shaves, especially the occipital area of the scalp.
- Avoid skin-irritating things like tight helmets, tight shirt collars, and wooden combs.
- Practice safer shaving habits to prevent the spread of blood-borne infections.

Homoeopathy a unique and holistic system of medicine [13]; it considers the patient as a whole rather than just treating the parts affected with its Individualistic approach towards the patient. This case report suggests an effective treatment measure for AKN.

Case Report

A fourteen-year-old male Christian student visited the OPD (January 22 2021) of the National Homoeopathy Research Institute in Mental Health, Under Central Council for Research in Homoeopathy, Kottayam, Kerala, presented papular, pustular eruption on the occipital region for six

months (Figure2). The lesions exuded the discharges on scratching; discharges were bloody, sometimes watery discharges, itching aggravating when touching the lesions, night, scratching. The patient was healthy six months back and assumed that these eruptions start after self-hair cutting with a trimmer during the pandemic lockdown.

He had a History of frequent upper respiratory tract infection, taking modern conventional medicine whenever needed without any adverse events.

In family history, his father is hypertensive, and his mother has had bronchial asthma; both are under modern conventional medicine.

On General Examination, height-138cm, weight, 44kg, BMI – 23.1kg/m²(overweight), and no other significant findings were noted in general and other systemic examinations. Local examination- eruptions- popular, pustular, grouped, on occipital region, no scales and fissures.

Homoeopathic Generals

Mental generals

After detailed case taking, he had a confusion of mind, especially while writing, making mistakes, the sadness of his appearance, fear of bad news, aversion to doing his regular work, and desire to travel.

Physical generals

The patient is fat and flabby, non-vegetarian with a good appetite, desire for meat, salty and spicy food like Lays, thirstless, perspiration more on scalp, thermally chilly person.

Analysis of the case

After analysis and evaluation, the characteristic symptoms were considered for constructing the totality like the confusion of mind, mistakes while writing, sadness, fear of bad news, indolence to work, desire to travel, desire to meat, salty and spicy things, thirstless, tendency to take cold easily, eruptions occipital region, cicatrices, itching aggravated at night, scratching, perspiration more on scalp and Obesity.

Miasmatic analysis of all the symptoms [Table 1] was done using Dr R.P Patel [14] Chronic Miasms in Homoeopathy and their cure, and the dominant Miasm was Psora.

The remedy selection was based on the repertorisation of the case using Synthesis Treasure edition 2009v (Schroyans. F), Radaropus2.2.16, - licence:121347. The repertorisation chart is shown in Figure:4

Table 1: Miasmatic Evaluation Of The Symptoms/ Rubric

Section/Symptoms/Rubric	Miasms and Marks			Pg. no
	Psora	Sycosis	Syphilis	
Mind-Confusion of mind	1	1		11
Mind-Fear of bad news	1			43
Mind-Indolence to work,	1			54
Mind-Mistakes while writing in		1		63
Mind-Sadness,	1		1	71
Mind-Travel desire to	1			82
Stomach-Desire to meat,	1			440
Stomach-Desire to salty	1		1	440
Stomach-Desire to spicy things,	1			440
Stomach- thirstless	1			477
Generalities-cold -tendency to take	1		1	1231
Head-eruptions occipital region	1			106

Head- perspiration- scalp		1	1	204
Skin-cicatrices,	1			1205
Skin-Itching aggravated at night	1			1220
Skin-Itching aggravated by scratching.	1			1220
Generalities-obesity		1		1244
Total	14	4	4	

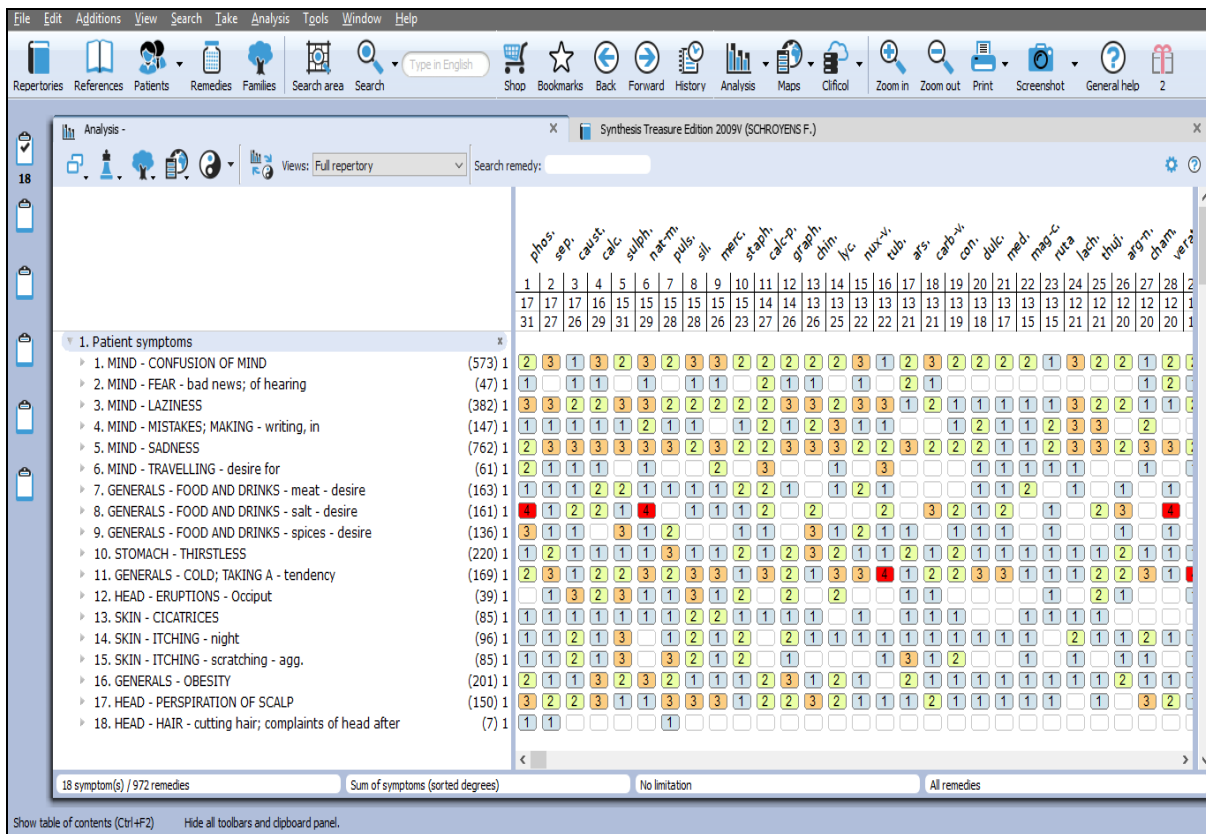


Fig 4: Repertorisation chart

Based on the repertorial analysis, two remedies (*Phosphorus* and *Calcarea carbnoica*) are coming close to the constitution; hence we followed the principles of the synthetic prescription ¹⁵and selected the *Calcarea*

Phosphoricum 30c four doses every 15 days once on an empty stomach morning followed by placebo for 60 days on the first visit due to corona pandemic and follow-up of the case with treatment [Table 2].

Table 2: Follow-up of the case with treatment

Date and visit	Sign and symptoms	Prescription and remarks
22.01.2021 First visit	Eruptions on the occipital region with itching and mild serosanguinous and bloody discharges occasionally.	1. Calcarea Phosphoricum 30c- four doses- 15 days once 2. Blank Tablets- bds/60 days.
23.04. 2021 Second visit	Mild decrease in the eruptions and no itching. No discharges. Still, the symptoms are present without any notifying new symptoms.	1. Calcarea Phosphoricum 30c- 2 doses. Monthly once days once. 2. Blank Tablets- bds/60 days
05.05. 2021 Third visit	Much improved in decreasing the size of eruptions without any discharges. Figure:3 Generals were good. No other complaints noticed.	1. Saclac 2 doses. Monthly once days once. 2. Blank Tablets- bds/60 days

Discussion

The above case report shows that constitutional prescription will helpful in the treatment of AKN. On repertorisation, *Phosphorus* came on hierarchy and but the physical constitution resembles *Calcarea carbonica*; hence, In this case, *Calcarea Phosphoricum* 30c was prescribed, which covered the physical constitution and repertorial totality, and it has shown optimistic results.

In this case, the total score of the outcome as per the changed Monarch Inventory ^[16] (Improved version of the modified Naranjo criteria for Homoeopathy was 8, which was close to the maximum score of 13 [Table 3]. Thus,

these results explicitly show the casual attribution of the single constitutional Homoeopathic treatment *Calcarea Phosphorus* towards the remission of the AKN in this case. The general concept in Homoeopathy towards treatment is 'treat the patient, not the disease'. In a superficial sense, this may be preposterous, but it conveys its holistic concept towards the patient in a more profound sense, which Kent J.T ^[17] explained in his lectures.

This concept is very similar to the Father of Modern Medicine; Sir William Osler says, "The good physician treats the disease; the great physician treats the patient who has the disease." ^[18].

In this case, the essential mental and physical generals and particulars, i.e., ailments after hair cutting, confusion of mind, fear of hearing bad news, indolence to work, mistakes while writing in, sadness, travel desire to, desire to meat, salty, spicy things, thirstless, tendency to take cold, eruptions occipital region, perspiration- scalp, Skin-cicatrices, Itching aggravated at night, by scratching, Obesity were included for repertorisation purpose.

There is no specific conventional treatment for AKN, and available treatment includes topical or oral antibiotics, retinoids, corticosteroids, topical fusidic acid and topical urea, intralesional injection of triamcinolone, and surgical excision, electrosurgery, cryotherapy, light and laser therapy. A combination of treatments may need, but they may not offer complete alleviation from symptoms of this illness.

In Homoeopathy, the constitutional prescription will help in treating and managing this rare and stubborn case. Furthermore, this intervention may arrest the development of cicatricial alopecia, which is the natural course and complication of the illness. First, however, a prospective research study is suggested for scientific validation as this is a single case report.

Conclusion

This present case report shows the role of Homoeopathy in treating and managing the cases of rare disease like AKN, where *Calcarea phosphoricum* was prescribed as a constitutional medicine showed evident positive results. This case report will enhance the clinicians' knowledge in suggesting proper management, which will benefit the patients suffering from AKN and similar rare disorders.

Declaration of the patient consent

The authors certify that the patient completed a consent form in which he gave his consent for his medical images and other clinical details to be reported anonymously in an academic journal. The patient understands that his name and initials will not be published and that all due efforts conceal his identity.

Acknowledgement

The authors acknowledge Dr. K.C. Muraleedharan, Officer in Charge, National Homoeopathy Research Institute in Mental Health, Kottayam, Kerala, for his encouragement and guidance regarding case report writing and scientific publication.

Table 3: Monarch Inventory (Improved version of the modified Naranjo criteria for Homoeopathy)

Domains	Yes	No	Not Sure or N/A	The score for successfully treated cases	Justification
Was there an improvement in the main symptom or condition for which the homoeopathic medicine was prescribed?	+2	-1	0	2	The patient came with eruptions on an occipital region with itching and serosanguinous and bloody discharges occasionally.
Did the clinical improvement occur within a plausible Timeframe relative to the medicine intake?	+1	-2	0	1	The patient had the complaint for six months before starting the medicine, and marked improvement seen in the first follow up itself.
Was there a homoeopathic aggravation of symptoms?	+1	0	0	0	Not observed
Did the effect encompass more than the main symptom or condition (i.e., were other symptoms, not related to the main presenting complaint, improved or changed)?	+1	0	0	1	Weight reduced from 44kg to 42kg even without modifications of the lifestyle.
Did overall well-being improve? (suggest using a validated scale or mention about changes in physical, emotional, and behavioural elements)	+1	0	0	1	Since the eruptions on the occipital region, it had affected his confidence which showed marked improvement after recovery.
Direction of cure: Did some symptoms improve in the opposite order of the development of symptoms of the disease?	+1	0	0	0	No
Direction of cure: Did at least one of the following aspects apply to the order of improvement in symptoms From organs of more importance to those of less importance? From deeper to more superficial aspects of the individual? From the top downward?	+1	0	0	0	Not applicable
Did 'old symptoms' (defined as nonseasonal and noncyclical symptoms that were previously thought to have resolved) reappear temporarily during the course of improvement?	+1	0	0	0	Not observed
Are there alternative causes (i.e. other than the medicine) that – with a high probability – could have produced the improvement? (consider known course of disease, other forms of treatment, and other clinically relevant interventions)	-3	+1	0	1	Not observed
Was the health improvement confirmed by any objective evidence? (e.g., investigations, clinical examination, etc.)	+2	0	0	2	Before and after photographs

Did repeat dosing, if conducted, create similar clinical improvement?	+1	0	0	0	Not sure.
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Fig 2: Before Treatment



Fig 3: After Treatment

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