22-Year-Old Male with Firm, Itchy Papules, and Hair Loss on the Occipital Scalp

15

Ryan Jay, Austin Cusick, Suchita Sampath, and Shannon C. Trotter

Abstract

Acne keloidalis nuchae (AKN) is a rare cicatricial alopecia that typically affects young African-American males. While the pathophysiology is not well described, there are several competing theories to explain the phenomenon including foreign body reactions to hair, lichen simplex chronicus, or a subtype of acne mechanica. The most accepted pathophysiology involves chronic folliculitis in genetically susceptible patients that is exacerbated by excoriations and continued inflammation. Diagnosis of AKN is usually clinical, and presentation is often described in stages from the nape of the neck to the vertex of the scalp. Physical exam will reveal papules and pustules of various sizes that may coalesce into plaques. These lesions can be accompanied by excoriations. There may also be overlying signs of bacterial infection secondary to trauma. Severe manifestations will be characterized by large smooth plaques that extend beyond the borders of the original pathology. While not required, dermoscopy and histopathology can help diagnose AKN. No standardized treatment regimen currently exists for AKN. The primary focus should be placed on preventing further trauma to pre-existing lesions. Moreover, antibacterial cleaners can be useful in reducing bacterial burden and subsequent infection. In early stages, treatment can begin with topical corticosteroids. Intralesional triamcinolone can be used for larger papules and plaques. Other potential medications include topical retinoids, topical clindamycin, or systemic tetracyclines. AKN characterized as "tumor stage" may

R. Jay (✉) · A. Cusick
OhioHealth Riverside Methodist Hospital, Columbus, OH, USA

S. Sampath
Ohio University Heritage College of Osteopathic Medicine, Dublin, OH, USA

S. C. Trotter
Dermatologists of Central States, Canal Winchester, OH, USA
e-mail: strotter@docsdermgroup.com

S. C. Trotter, S. Sampath (eds.), *Clinical Cases in Alopecia*, Clinical Cases in Dermatology, https://doi.org/10.1007/978-3-031-15820-9_15

require surgical intervention, and several approaches exist. The primarily described approach includes horizontal elliptical excision with healing by secondary intention. This treatment has been expanded upon to include possible staged excisions utilizing skin grafts or varying excision approaches including the occipital protuberance. Breakthrough papule formation can be treated with high potency topical corticosteroids or intralesional steroids. Treatment of AKN largely depends on the physical presentation and desired cosmesis.

Keywords

Acne keloidalis nuchae · Cicatricial alopecia · Chronic folliculitis · Spade sign

A 22-year-old male presented with several hard papules on the occipital scalp of 9 months duration that started after he entered basic training for the army. He admitted to scratching at the lesions constantly. Some of the papules seemed to come and go, but most were constant and lacked hair growth.

On physical examination, there were firm papules and areas of scarring alopecia noted on the occipital scalp with few scattered pustules (Fig. 15.1). The rest of the scalp was spared. The eyebrows and eyelashes were of normal density. The fingernails were normal.

Based on the clinical case description, what is the most likely diagnosis?

1. Tinea capitis
2. Dissecting cellulitis
3. Acne keloidalis nuchae
4. Folliculitis decalvans

Fig. 15.1 Areas of scarring alopecia along with scattered pustules and multiple firm papules along the occipital scalp

Diagnosis

Acne keloidalis nuchae.

Discussion

Acne keloidalis nuchae (AKN) is a rare and chronic scarring (cicatricial) alopecia involving the hair follicles that predominantly affects post-pubertal men of African descent under 55 years of age [1]. The incidence ranges between 0.45% and 9%, with most cases in populations with darker skin and curlier hair [2]. Risk factors and associated conditions linked to AKN include obesity, hypertension, chronic scalp folliculitis, pseudofolliculitis barbae, increased frequency of hairdressing, and close hair cropping [3, 4].

Though the name suggests it encompasses keloids or acne, AKN is not characterized by true forms of either one. The pathophysiology is not entirely known and may be multifactorial, but several hypotheses have been postulated without significant support. These include AKN as a transepithelial elimination disorder stemming from a foreign body reaction to misplaced hair, lichen simplex chronicus with scarring, a foreign body reaction to ingrown hairs, and a variation of acne mechanica. The more widely accepted pathophysiology is chronic folliculitis from rubbing, scratching, or irritation to the occiput and nape that leads to scarring [5]. Diagnosis is usually clinical, and the presentation may appear variably based on the stage of lesions. Often preceded by pruritus, the onset of AKN presents anywhere from the nape of the neck to the vertex of the scalp with various-sized papules or pustules that may coalesce. Active areas may bleed, itch, or feel painful. Subsequent manipulation by the patient could induce a secondary bacterial infection. Chronic irritation then leads to keloid-like nodules and plaques with alopecia [1] (Fig. 15.2). Dermoscopic findings may include perifollicular pustules, perifollicular fibrosis, or tufted hairs are found [6]. Histopathology does not offer specific findings outside of

Fig. 15.2 Various sized papules and pustules along the nape of the neck. Image courtesy of Dr. Melissa Piliang and Janine Sot, MBA medical photographer

features common to other scarring alopecia, such as perifollicular inflammation with lymphocytes and plasma cells, peri-isthmic lamellar fibrosis, and absent sebaceous glands [7]. However, recently, a pathognomonic finding during the subacute stage of AKN was proposed [8]. The finding is called a "spade sign" and describes the inflammation leading to a dilated spade-like space in the lower isthmus of hair follicles [8].

Treatment

AKN is difficult to manage because no widely accepted treatment regimen exists. Numerous measures from case series have reportedly helped with varying degrees of success. Efficacy of treatment depends upon the stage and extension of lesions, with multimodal approaches often working the best. First and foremost, preventative measures must be taken to combat mechanical irritation. This includes wearing non-irritative clothing or collarless shirts and avoiding close shaving of the occiput and nape of the neck. Antibacterial cleaners may also be implemented in the treatment of secondary infections [2]. In the earlier stages where only small papules and no keloid-like scars are present, treatment often begins with topical corticosteroids to the affected region and intralesional triamcinolone for larger papules and plaques [1]. Simultaneous topical retinoids for mild disease or topical clindamycin for pustular presentations may be utilized with corticosteroids. In more extensive mild-to-moderate cases, systemic tetracyclines may also be used for anti-inflammatory and antibacterial properties [9].

Severe or later-stage cases that are resistant to medical therapies and present with large keloid-like nodules and plaques may consider surgery. Surgical excision with a horizontal ellipse involving the posterior hairline and healed by secondary intention led to resolution and favorable cosmetic results [10, 11] (Fig. 15.3). However, others found success with a multi-faceted surgical technique. Galarza et al. treated giant 'tumor stage' AKN with radical surgical excision and initial secondary intention healing for 50 days, which allowed the wound to contract and heal before utilizing a skin graft [12]. In an intervention before-after trial of 25 patients undergoing surgical excision for refractory AKN, little risk of recurrence was found for cases with primary closure (though not as favorable in cosmesis), and those with especially large lesions responded well to a staged-excision approach [13]. An innovative and successful surgical method by Umar et al. utilized a bat-shaped excision of affected areas confined to the occipital protuberance superiorly and the posterior hairline inferiorly, followed by secondary intention healing with or without tension suture assistance [14]. If minimal recurrence of papules or pustules happened after surgical excision, high potency topical steroids and intralesional steroids usually resolved them [13]. Overall, providers should decide on a surgical approach based on the severity of the case and desired cosmetic outcome.

Laser hair removal represents another potential treatment for AKN. Long-pulse, long-wavelength lasers are preferred because they can better penetrate the

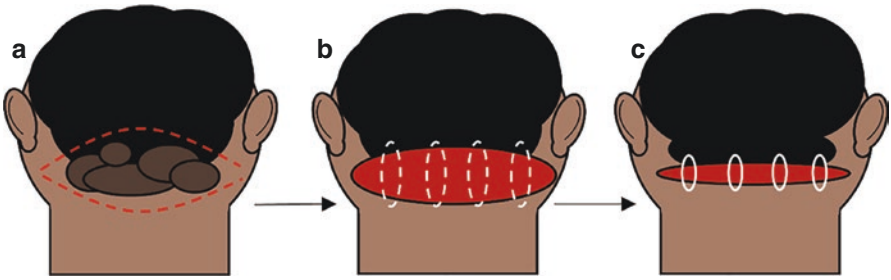

Fig. 15.3 Surgical procedure with a horizontal ellipse for severe AKN. Panel **a** depicts AKN and the red dotted line depicts an elliptical excision pattern. Panel **b** demonstrates the surgical defect and placement of tension sutures (white dotted line). Panel **c** shows the final product of tension sutures with partial closure allowing the remaining defect to close by secondary intention

2–4 mm average depth of follicles [14]. 1064 nm Nd-YAG may theoretically work the best because it has the deepest penetration while damaging melanocytes the least [15]. A comparative study between Er:YAG and Nd:YAG lasers found that both led to a significant decrease in the number of papules, as well as the size and consistency of plaques [16]. Similarly, a study with 17 patients utilized 6 sessions of 755 nm alexandrite laser therapy and demonstrated a significant improvement in mean papule/pustule count, plaque size, and pliability of lesions [17]. A combination of lasers with other treatment modalities, such as Nd-YAG with topical corticosteroids or excision followed by Nd-YAG, can be personalized for severity, and may provide a more favorable outcome [18, 19]. For those who are neither surgical nor laser candidates, or have remained refractory to all the aforementioned treatments, low-dose radiotherapy may lead to resolution [20, 21].

Treatment of AKN depends on the type, size, and extent of lesions, and multimodal approaches are often needed. Patients should also be counseled on the difficulties of AKN management and treatment. Initially, preventative measures should be implemented followed by progressively invasive therapies as needed.

Key Points

- Acne keloidalis nuchae (AKN) tends to affect post-pubertal men of African descent with curly hair, though other demographics can be afflicted too.
- Early lesions can present on the occiput and nape as small papules and pustules, but later stages present as larger keloid-like nodules and plaques.
- Treatment begins with topical and intralesional corticosteroids with or without systemic tetracyclines, while later staged or severe disease may require surgery.
- Laser hair removal done at earlier stages may be an effective treatment that prevents future occurrences.

References

1. Ogunbiyi A. Acne keloidalis nuchae: prevalence, impact, and management challenges. Clin Cosmet Investig Dermatol. 2016;9:483–9. https://doi.org/10.2147/CCID.S99225.
2. Maranda EL, Simmons BJ, Nguyen AH, Lim VM, Keri JE. Treatment of Acne keloidalis nuchae: a systematic review of the literature. Dermatol Ther (Heidelb). 2016;6(3):363–78. https://doi.org/10.1007/s13555-016-0134-5.
3. Saka B, Akakpo AS, Téclessou JN, Mouhari-Toure A, Kassang P, Gnossike P, et al. Facteurs de risque associés aux folliculites fibrosantes de la nuque chez le sujet noir : étude cas-témoins [Risk factors associated with acne keloidalis nuchae, in black subjects: A case-control study]. Ann Dermatol Venereol. 2020;147(5):350–4. https://doi.org/10.1016/j.annder.2020.01.007.
4. East-Innis ADC, Stylianou K, Paolino A, Ho JD. Acne keloidalis nuchae: risk factors and associated disorders—a retrospective study. Int J Dermatol. 2017;56(8):828–32. https://doi.org/10.1111/ijd.13678.
5. Shapero J, Shapero H. Acne keloidalis nuchae is scar and keloid formation secondary to mechanically induced folliculitis. J Cutan Med Surg. 2011;15(4):238–40. https://doi.org/10.2310/7750.2011.1005.
6. Chouk C, Litaiem N, Jones M, Zeglaoui F. Acne keloidalis nuchae: clinical and dermoscopic features. BMJ Case Rep. 2017;2017:bcr2017222222. https://doi.org/10.1136/bcr-2017-222222.
7. Doche I, Coelho EQ, Quaresma MV, da Matta Rivitti-Machado MC. Acne keloidalis nuchae and folliculitis decalvans: same process affecting the follicle or coexisting diseases? A retrospective study. Int J Dermatol. 2019;58(10):e200–3. https://doi.org/10.1111/ijd.14565.
8. Cheng AY, Lee CN, Hsieh FN, et al. "Spade sign" and inflammation/fibrosis limited to the upper and mid-dermis as the pathognomonic features of acne keloidalis. J Dermatol. 2020;47(1):41–6. https://doi.org/10.1111/1346-8138.15127.
9. Alexis A, Heath CR, Halder RM. Folliculitis keloidalis nuchae and pseudofolliculitis barbae: are prevention and effective treatment within reach? Dermatol Clin. 2014;32(2):183–91. https://doi.org/10.1016/j.det.2013.12.001.
10. Glenn MJ, Bennett RG, Kelly AP. Acne keloidalis nuchae: treatment with excision and second-intention healing. J Am Acad Dermatol. 1995;33(2 Pt 1):243–6. https://doi.org/10.1016/0190-9622(95)90242-2.
11. Bajaj V, Langtry JA. Surgical excision of acne keloidalis nuchae with secondary intention healing. Clin Exp Dermatol. 2008;33(1):53–5. https://doi.org/10.1111/j.1365-2230.2007.02549.x.
12. Galarza LI, Azar CA, Al Hmada Y, Medina A. Surgical management of giant acne keloidalis nuchae lesions. Case Reports Plast Surg Hand Surg. 2021;8(1):145–52. Published 2021 Sep 23. https://doi.org/10.1080/23320885.2021.1982392.
13. Gloster HM Jr. The surgical management of extensive cases of acne keloidalis nuchae. Arch Dermatol. 2000;136(11):1376–9. https://doi.org/10.1001/archderm.136.11.1376.
14. Umar S, David CV, Castillo JR, Queller J, Sandhu S. Innovative surgical approaches and selection criteria of large acne keloidalis nuchae lesions. Plast Reconstr Surg Glob Open. 2019;7(5):e2215. https://doi.org/10.1097/GOX.0000000000002215.
15. Umar S. Selection criteria and techniques for improved cosmesis and predictable outcomes in laser hair removal treatment of acne keloidalis nuchae. JAAD Case Rep. 2019;5(6):529–34. https://doi.org/10.1016/j.jdcr.2019.02.034. [published correction appears in JAAD Case Rep. 2019 Aug 29;5(9):809]
16. Gamil HD, Khater EM, Khattab FM, Khalil MA. Successful treatment of acne keloidalis nuchae with erbium:YAG laser: a comparative study. J Cosmet Laser Ther. 2018;20(7–8):419–23. https://doi.org/10.1080/14764172.2018.1455982.
17. Tawfik A, Osman MA, Rashwan I. A novel treatment of acne keloidalis nuchae by long-pulsed alexandrite laser. Dermatol Surg. 2018;44(3):413–20. https://doi.org/10.1097/DSS.0000000000001336.

18. Woo DK, Treyger G, Henderson M, Huggins RH, Jackson-Richards D, Hamzavi I. Prospective controlled trial for the treatment of acne keloidalis nuchae with a long-pulsed neodymium-doped yttrium-Aluminum-garnet laser. J Cutan Med Surg. 2018;22(2):236–8. https://doi.org/10.1177/1203475417739846.
19. Madura C, Kareddy S, Kusuma MR, Chandrashekar BS. Multimodality surgical treatment approach to acne keloidalis nuchae based on lesion morphology. J Cutan Aesthet Surg. 2021;14(2):256–9. https://doi.org/10.4103/JCAS.JCAS_32_20.
20. Millán-Cayetano JF, Repiso-Jiménez JB, Del Boz J, de Troya-Martín M. Refractory acne keloidalis nuchae treated with radiotherapy. Australas J Dermatol. 2017;58(1):e11–3. https://doi.org/10.1111/ajd.12380.
21. Umar S, Sila CR. Acne keloidalis nuchae: a role for low-dose radiotherapy. JAAD Case Rep. 2021;13:90–3. https://doi.org/10.1016/j.jdcr.2021.05.008.