Acne Keloidalis Nuchae – Overlooked and Undertreated?

SONDRE FLAGSTAD KOREN, ANNE KATRINE LORENTZEN AND CAMILLA ASKLUND

Department of Plastic Surgery, Herlev and Gentofte Hospital, Copenhagen University, Borgmester Ib Juuls Vej 5, DK-2730 Herlev, Denmark. E-mail: sondre.flagstad.koren@regionh.dk

INTRODUCTION

Acne keloidalis nuchae (AKN), also known as alopecia cicatrisata/folliculitis keloidalis nuchae, is a chronic inflammatory disease of the occipital/nuchal hair follicles. It typically debuts after puberty and untreated it may lead to pruritus, pain, keloid-like scar formation of the occipital region and unsatisfying cosmesis. Mechanical manipulation of affected areas from scratching leads to further inflammation and disease progression. AKN primarily affects people of darker skin, and men are affected 20 times more frequently than women (1). A study examining the spectrum of skin diseases in a black population in south-east London, referred to a dermatology clinic by their general practitioner, found AKN as the diagnosis in 13.7% of the patients (2). AKN is to our knowledge not described in Nordic literature.

Clinically AKN presents with inflamed papules with peripapular erythema in the neck that can develop into fibrosis and further coalesce into hairless plaques and noduli. The lesions predispose for local infection. The pathogenesis is unknown, but hormonal imbalance with excess of androgens and trauma/irritation have been proposed as aetiological factors (3).

In early stages, AKN may be treated medically, but later stages require laser therapy or surgery.

CASE

A 45-year-old man, born in Nigeria, presented with a 4 × 2 cm fibrotic, keloid like plaque in the occipital region (Fig. 1A). The patient had been having painful, pruritic occipital papules for several years, and surgical excision had been performed in Nigeria years earlier. The lesion had now recurred, and two other minor lesions resembling the primary lesion were seen in close proximity. Pruritus was the primary symptom. We performed an intralesional excision, the traditional Danish plastic surgery approach to excision of keloids, and histology showed folliculitis with active and chronic inflammation and fibrosis. Folliculitis keloidalis was proposed, and as this was in accordance with the clinical presentation, a diagnosis of AKN was made (Fig 1B). Postoperatively the patient was given a total of 6 injections of intralesional glucocorticoids, with an interval of 3–4 weeks. This resulted in a reduction of the size of the lesion, as well as alleviation of the pruritus. Ten months later, there were recurrent symptoms of occipital pruritus and pain (Fig 1C), and the lesion was again treated with surgical intralesional excision (Fig. 2). One month after the operation, the patient complained that the lesion was recurring, as was the pruritus. The lesion was treated with 3 intralesional glu-
cocorticoid injections, over the following 10 weeks, and the patient reported effect on the pruritus (Fig 1D). He did not attend further follow-up appointments after the last injection.

**Discussion**

The patient in this case presented with late stage AKN with classic keloid-like scar formation in the occipital region. The disease can be treated by avoiding mechanical irritation such as scratching, and avoiding clothing/helmets that rub against the papules. Avoidance of this alone has shown effect where previous medical treatment had failed (4).

Systemic treatment with retinoids and antibiotics – used as monotherapy or in combination with topical antibiotics – have shown effect on the size of the papules in the neck. The treatment may last for up to a year, and there is risk of recurrence after the treatment is discontinued. Intralesional injections with glucocorticoids have shown effect and can be combined with antibiotics and/or retinoids (1).

Laser treatment with 1064-Nd:YAG laser once per month for 5 months, showed a mean effect of 82%, when looking at the reduction in size and numbers of the papules. There was a significantly greater effect when treatment was initiated early (5). A significant decrease in the number and size of plaques was also seen when AKN was treated with Er:YAG laser (6). Surgical excision is recommended for late stage AKN with fibrotic plaque formation. The most effective treatment with optimal cosmetic result is a deep excision in unaffected tissue around the lesion (opposed to the traditional Danish plastic surgical approach to keloid treatment, where the strategy typically is intralesional excision as a single procedure or serial excision) and primary closure or second intention healing (1, 7).

The patient was treated in our department, a specialized centre. The lesion clinically resembled a typical keloid, and was therefore treated as such. We believe the fact that the lesion was still treated with intralesional excisions shows that there is a potential risk that other patients do not receive the recommended extralesional deep excision either. Further knowledge about the disease, and a preoperative punch biopsy could potentially have changed our treatment strategy.

AKN is to our knowledge previously undescribed in Nordic literature, but with an ongoing globalization, the patient group may be overlooked and undertreated. Our goal with this case report is to help clinicians make an early diagnosis so that the patients with early stage AKN may be treated medically, and so that patients with later stage AKN may receive the recommended surgical treatment.

**Conflict of interest**

The authors declare that they have no conflicts of interest.

**References**


**Fig 2.** Intralesional excision, intraoperative picture from the second operation.