Acne keloidalis nuchae: A role for low-dose radiotherapy



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Key words: acne keloidalis nuchae; low-dose; outcome; radiation; radiotherapy; treatment.

INTRODUCTION

Acne keloidalis nuchae (AKN) is a folliculocentric scalp disease mainly affecting men of color. Numerous studies have shown that it is not a true keloid¹⁻³; when all follicular structures within the lesion are eliminated by surgery or laser, the lesion is permanently eliminated.³⁻⁶ Millan-Cayetano et al⁷ reported the use of radiation therapy (RT) in a patient with an isolated AKN lesion with no recurrence. Permanent killing of lesional hair by radiation coincided with lesion regression and permanent elimination of the disease, further supporting the folliculocentric foundation of AKN, which is resolved by focusing the treatment objective on removing hair located within the lesion.⁷

We describe the use of low-dose RT to cure AKN in a patient who failed years of medical therapy and who was not a candidate for primary surgical intervention.

CASE REPORT

A 39-year-old Pacific-Asian-American man developed AKN beginning in the nape of his head at the age of 10 with subsequent spread to his entire scalp. This spread coincided with the development of excessive scalp folds—cutis verticis gyrata (CVG). His symptoms included significant pain, swelling, and discharge of pus and blood. Examination revealed inflamed plaques with tufted hair involving most of the scalp, sparing all CVG fold recesses (Fig 1, *A*) except the inferior occipital CVG fold, which exhibited multiple fibrotic-looking AKN papules (Fig 1, *C*). He had bitemporal AKN masses with a discharging sinus on the left temporal mass (Fig 1, *B*). Additionally, he had severe cystic acne, rhinophyma, and pseudofolliculitis barbae. We performed Abbreviations used:

- AKN: acne keloidalis nuchae
- CVG: cutis verticis gyrata
- RT: radiation therapy

additional tests (X-rays and insulin growth factor level and thyroid function tests) to rule out pachydermoperiostosis, acromegaly, and thyroid disease. A bacterial culture of purulent discharge revealed *Staphylococcus aureus*, for which the patient was treated with ciprofloxacin.

Past treatments with isotretinoin, colchicine, finasteride, topical and oral antibiotics, and intralesional and topical steroids were unsuccessful. He was considered a poor candidate for surgical removal of individual lesions because of widespread disease. Total scalp excision followed by skin grafting after a long granulation period was considered but abandoned because of his poorly controlled diabetes, hypertension, and obesity. Laser hair removal was not an option due to the thickness of the plaques and masses, which would have limited the ability of laser treatment to reach the follicular killing zones of the bulge (where stem cells reside) and bulbs (where the dermal papillae cells reside).⁵ For these reasons, we chose RT.

Our primary goal was to minimize the amount of radiation used by selecting the amount sufficient to kill intralesional hair (20 sessions at 2 Gy per session at 6 MeV). Furthermore, minimal-dose RT is used as a tool to rapidly ameliorate the signs and symptoms of inflammation, since studies have documented a robust response of inflammatory conditions to low-dose radiation.⁸

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Fig 1. Widespread AKN prior to radiotherapy. **A**, Posterior view showing an inflamed plaque with tufted hairs with sparing of superior occipital CVG fold, which shows the presence of normal hair (*green arrows*). The inferior CVG fold exhibits fibrotic-looking AKN papules with tufted hairs (*yellow arrows* seen also in B and C). **B**, Side view showing an inflamed mass in the left temporal area, which has a discharging sinus. CVG folds in the parietal area exhibit normal hair growth with absence of disease (*green arrow*). Inferior CVG fold, where AKN papules exist (*yellow arrows*) **C**, close-up view of the nape showing the inferior CVG fold, which is less inflamed and showing multiple merging AKN papules and tufted hair (*yellow arrows*). *CVG*, Cutis verticis gyrate.

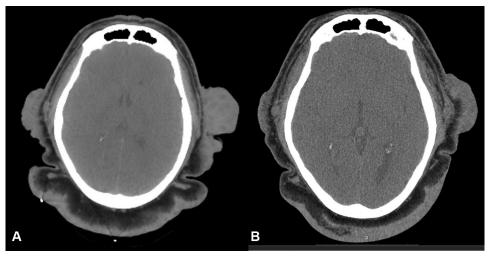


Fig 2. Non-contrast computed tomography scans of the head, coronal sections. **A**, Prior to radiotherapy showing significant subcutaneous tissue volume. **B**, 14 months post-radiotherapy, showing overall reduction in subcutaneous tissue volume in affected areas.

Volumetric-modulated arc therapy allowed us to treat the entire scalp while significantly minimizing underlying brain exposure and sparing the adjacent parotid glands. A custom 1-cm bolus was placed over the entire area to ensure a full dose to the thickness of the lesion, including the hair follicle. After treatment, the patient experienced dysgeusia and mild xerostomia, which resolved.

Symptoms of pain, swelling, and pus discharge disappeared halfway through the RT course. Within 3 days of treatment, the patient had lost all his hair. However, hairs in the clinically less-inflamed inferior occipital CVG groove, where fibrotic AKN papules resided, as well as all non-disease areas (all remaining CVG folds), returned within 5-6 months, while all hairs in areas of clinically inflamed AKN plaques and masses were lost permanently (Fig 3). The permanent elimination of intralesional hair coincided with permanent clearance of AKN lesions and gradual regression of tissue volume in all the affected areas of the scalp, including all the plaques and masses (Figs 2 and 3). We noted, however, the persistence of a thin serous discharge on the original sinus of the left temporal mass. We also noted the persistence of the fibrotic AKN papules in the clinically less-inflamed recesses of the inferior occipital CVG area, where

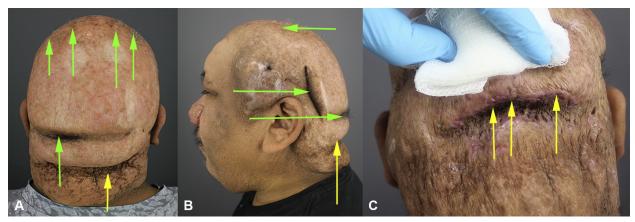


Fig 3. Ten months after radiotherapy. **A**, Posterior view. All inflammation had resolved, all plaques recessed in volume, and all hair eliminated permanently, except for hairs in the non-inflamed (all CVG folds, *green arrows*) and less-inflamed recesses of the inferior occipital CVG groove (*yellow arrows*). **B**, Side view showing elimination of inflammation in plaques and masses, with persistent sinus discharge from the left temporal mass. Permanent radiation-induced alopecia was achieved in the entire scalp, except in the non-inflamed areas (all CVG folds, *green arrows*), including and the inferior occipital CVG groove (*yellow arrow*). **C**, Nape showing the inferior occipital CVG fold with persistent hair and AKN papules (*yellow arrows*). *CVG*, Cutis verticis gyrate.



Fig 4. Four months after surgery (about 10 months after radiotherapy). Surgery involved removal of the left temporal mass, debulking of the lower occipital plaque, and removal of recalcitrant AKN and the inferior occipital CVG fold containing it. Lesions were excised and allowed to close by second-intention healing guided by tension sutures. Cosmetic improvement was noted as well as uneventful post-surgical wound healing in these radiation-treated areas. **A**, Posterior view showing elimination of the inferior occipital CVG fold and AKN lesions and hair that had survived low-dose radiation. **B**, Side view demonstrating the left temporal area and occipital area with a good healing outcome after removal of the left temporal mass and inferior occipital AKN-involved CVG fold. A disease-free hair-containing CVG fold of the left parietal area is also shown. *CVG*, Cutis verticis gyrate.

intralesional hair had remained (Fig 3, *C*). Histologic examination of tissue taken from 2 areas confirmed dense and well-organized scarring in the less

responsive areas compared with the loose and disorganized fibrosis found in the rest of the affected zones where complete disease remission occurred in response to RT. The left temporal mass and the inferior occipital CVG folds were subsequently excised with complete and uneventful wound healing and absent AKN disease recurrence (Fig 4). Sixteen months after treatment, the patient is completely disease-free in the areas that responded, and the treated area continues to improve, as the very thick skin he had is returning closer to normal.

DISCUSSION

In the present case, surgery alone would have involved total scalp excision followed by skin grafting after a long period of allowing granulation tissue to grow in. Given the patient's history of diabetes, high blood pressure, and significant obesity, the surgery would have carried a significant risk, and the healing course would entail a high risk of complications.

The criteria for treating widespread AKN lesions with RT in our patient were the failure of 14 years of conservative medical therapies, the unfeasibility of surgery, and the non-viability of hair elimination by laser because of the excessive depth of the follicle bulge and root.⁵ Both the cumulative dose and the fractional dose used are important in causing alopecia with radiotherapy.⁹ Our experience indicates that the radiation dose needed to permanently kill hair and eliminate the disease in inflamed and friable AKN lesions is less than the dose required for non-inflamed areas of the scalp.

This offers the advantage of removing the need for high-radiation doses in AKN treatment and the advantage of selectively killing hair only in involved areas by using low-radiation doses. Patients should be fully informed about and consent to the risks and possible complications of RT.⁷ By avoiding high-radiation doses, complications such as swelling, telangiectasia, and skin cancer are reduced.^{10,11} It is expected that someone radiated at the age of 30 has an "excess absolute risk" of 44 in 10,000 for the development of skin cancer by the age of 70 for a dose of 40 Gy.¹¹

Ionizing radiation preferentially kills actively dividing cells at the time of mitosis. Slowly-dividing cells or cells in terminal stages of differentiation, such as densely organized scars of some AKN zones, can present areas of resistance to low-dose RT and may require secondary treatments with surgery. Using lower radiation doses, the tissue left in the wake of treatment is more likely to react well to surgeries with less chance of wound healing complications, as evidenced in our diabetic patient (Fig 4). AKN is not a keloid,^{1,2} so whereas RT is effective when applied to post-surgical excision of true keloids,¹² it

will generally be ineffective if used on nonsurgerized keloids.¹³ By contrast, in AKN, radiation is effective in non-surgerized keloids,⁵ with lowfractional doses minimizing wound healing risks from surgeries performed after RT.

CONCLUSION

RT can be used as a last resort to successfully treat debilitating AKN disease refractory to conservative treatments, while surgery and laser are either not feasible or viable.

Conflicts of interest

None disclosed.

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