Whole Scalp Radiation Therapy for Recurrent Benign Cylindromas

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Abstract
Cylindromas are a rare benign pathology that can manifest as multiple lesions on a patient's scalp. Standard of care is resection of lesions. We report a case of a 74-year-old man with a known diagnosis of Brooke-Spiegler syndrome which is a genetic syndrome that results in multiple recurrent cylindromas. He had approximately 70 prior resections to remove recurrent lesions with multiple grafts. After a large scalp recurrence, with multiple satellite areas, he preferred radiation to the largest site for no-surgical management. After an excellent clinical response, this led to him electing for total scalp irradiation to the remaining sites. He now has a complete response to all remaining sites.

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Introduction
Cylindromas are classified as benign neoplasms stemming from dermal lineage which rarely become malignant. Cylindromas present on faces and scalps as a slow-growing mass(es) and are more often found in females than in males [1]. Multiple cylindromas can occur in Brooke-Spiegler syndrome or familial cylindroma [2]. These cells appear to be apocrine in origin and have an association with spiradenomas. A mutation in the tumor suppressor CYLD gene results in uninhibited Wnt/B-catenin signaling which leads to unregulated growth [2, 3]. Surgical resection is the typical management though patients often present with recurrent lesions on the scalp leading to a turban appearance. Although benign, these lesions
often are morbid due to frequent bleeding, psychosocial stress, and painful requiring multiple surgeries [4]. We present a case report of a patient with Brooke-Spiegler syndrome who chose to undergo definitive radiation therapy for treatment of persistent scalp lesions after 70 recurrences causing extreme emotional distress and depression. His dermatologist thus recommended him to try radiation therapy given its utilization in uncontrolled spiradenomas.

**Case Presentation**

Mr. X is a 74-year-old man who presented to discuss nonoperative management for cylindromas on his scalp. The patient was diagnosed with Brooke-Spiegler syndrome in 1990 and since then has had recurrent cylindromas on his scalp. He has had over 70 surgical procedures performed in the past to remove the lesions on his head and now presented to attempt an alternate definitive management. He was referred by his dermatologist for evaluation, if radiation could help.

For most of his life, he would have resections on his scalp, sometimes followed by electrocautery. Frequently, he would have regrowth or a new area(s) within 6–8 weeks of each resection. Within the 8 months prior to presentation, he had a 5-cm lesion resected with a large graft. Histology of previously resected lesions demonstrated cells primarily of eccrine origin with a lack of lymphoid tissue suggesting cylindroma instead of a spiradenoma. A few months later, a new lesion grew lateral to the graft site and was enlarging.

The lesion that grew lateral to the graft was painful. He also presented with several painful lesions in the posterior auricular area. He also noted growth of a multitude of other lesions on his scalp which were new. He noted that some of the lesions hurt and bleed with minor irritation (i.e., his hat touching it or rolling on a pillow).

He wished to not receive further surgery and wanted radiation alone to manage his disease. He presented with diffuse lesions on his scalp. His most bothersome lesions were the primary mass, which measured about 5 × 4 × 2 cm, on his left scalp (Fig. 1) and a series of 3–4 small palpable nodules near his ear (Fig. 2).

The patient then elected to pursue a definitive radiation course to see if it would respond. Simulation CT scan was performed with 3-mm slices for radiation therapy treatment planning. Contouring of the scalp lesion was done based on the axial view. CT imaging helped focus on which regions to contour for the GTV, CTV, and PTV [5]. We considered the GTV to be the CTV for the primary mass and created a 5-mm expansion for PTV which we aimed to cover with a 3-D field. Photon therapy was chosen to treat the dominant lesion on the scalp. A half beam block (Fig. 3) was utilized to adequately target the tumor and minimize the normal tissue. The isocenter as depicted was placed in the bone, thus allowing adequate coverage of the lesion.

![Primary lesion on the scalp](image-url)
and minimizing divergence into the brain. The lesions on the patient’s temple and ear region were treated with electron therapy “en face.” This was contoured as a single CTV and treated with 2-cm margin for the electron field.

A 1-cm bolus was used as a tissue equivalent for adequate dose build-up on his scalp lesions and treated with 9E electrons. He was treated with 6,840 cGy in 38 fractions to both the dominant lesion as well as to the lesions on his lateral scalp.

The patient noted a decrease in the size of his lesion as well as less irritation of the smaller lesions near his ear. He had subjective improvement in size and firmness of lesion and was satisfied with his outcome. He had mild erythema and grade 1 dermatitis. At 4 weeks as per the patient, the dominant lesion had shrunk in size and continued to decrease in firmness. Measurement of the height revealed it to be around 4 mm compared to prior 2 cm. Given his marked improvement in the dominant lesions, he decided to have entire scalp treated a 4 months later to the same dose, 6,480 cGy, using an IMRT approach and avoiding the previously treated regions. His V60 (volume receiving 60 Gy) to the brain was 1.1%, and his V30 was 11%. His cochlea mean dose on the right and left were 1,163 cGy and 753 cGy, respectively, compared to an acceptable mean dose less than 4,500 cGy. He had a clinical complete response to his lesions and had minimal toxicity with some grade 2 dermatitis. His
Discussion/Conclusion

Cylindromas are an uncommon benign tumor often found on the scalp. Multiple cylindromas is a hallmark of Brooke-Spiegler syndrome, as seen in our patient, or in familial cylindromas [6]. The cell origin of cylindroma is suggested to be of both apocrine and eccrine origin [7]. The cylindrical morphology seen in Brooke-Spiegler syndrome supports eccrine origin; yet CD200, a hair stem cell marker, is present along with proteins linked to hair-specific differentiation such as KRT17. On physical examination, patients present with multiple smooth and firm tumors. Due to their location as the tumors enlarge, they can resemble headwear, hence the colloquial term “turban tumors” [8].

Due to the benign nature of these, the primary suggested therapeutic modality is surgery via either primary excision, curettage, electrosurgery, or laser resurfacing [9]. In fact, radiotherapy via speculation has not been recommended as first line due to concern over loss of a tumor suppressor gene CLYD1, which could induce further malignancy [9]. However, in other disease sites such as breast cancer, loss of important tumor suppressor genes such as ATM in it of itself is not a contraindication to receiving radiotherapy and does not demonstrate increase in normal tissue side effects [10]. Furthermore, despite basal cell carcinoma demonstrating loss of several tumor suppressor genes, including CLYD, radiation is still routinely used to manage certain lesions without increase in side effects [11].

Radiation has been discussed in the literature albeit sparsely. It has been discussed as playing a possible adjuvant role in the rare cases of malignant transformation of cylindromas [12]. Benign cylindromas have also been reported to be treated with radiofrequency ablation [9], but no evidence can be found of radiotherapy being done in the upfront setting in these cases and can currently be clearly identified in the literature.

Our patient underwent dozens of surgical procedures and opted for radiation therapy in lieu of additional resection. Surgical resection is the most used standard of care to remove these lesions but is often associated with a localized recurrence, especially with this genetic aberrancy.

When approaching this case, we viewed the histology of the patient’s cylindromas to be akin to desmoid tumors. Desmoid tumors are nonmalignant proliferations of fibroblasts that can respond to radiation therapy, albeit surgical resection is the first-line approach [13].

This case highlights several important points. The first is that if a certain treatment paradigm such as serial resections is not working, then it may be best to consider alternative approaches. Secondly, alternative approaches that are less invasive, such as radiation, offer...
similar success with proliferating cells, and despite lack of tumor suppressor genes does not preclude its use. With advanced techniques like IMRT the whole scalp can be targeted with acceptable low doses to the nearby brain. Given the sparsity in the literature, it was a reasonable approach to try definitive radiation to the dominant lesion and the small area in the auricular space which were symptomatic. Once they demonstrated response, a proper staged approach was able to be performed to irradiate all the remaining sites on the skull. Further follow-up will be performed on this case.

**Statement of Ethics**

Ethical approval was not required for this study in accordance with local/national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

**Conflict of Interest Statement**

The authors have no conflicts of interest to declare.

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**Author Contributions**

Shivam Kaushik is the medical student who wrote the report up and did the literature search for writing purposes. Neil Newman is the physician who treated the patient and assisted with editing and writing the manuscript. Danielle Siegel assisted with dosimetry and writing. Kenneth Niermann is the attending physician who directed Neil Newman and Shivam Kaushik throughout the manuscript development process.

**Data Availability Statement**

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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