http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v12i02.11

IGM Publication Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

A Rare Case of Proliferating Trichilemmal Tumor Treated with Radiotherapy

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Abstract
Proliferating Trichilemmal Tumor (PTT) is a rare tumor of the outer root sheath of the hair follicle with different biological behavior. They usually develop in older patients, most commonly affecting women, from pre-existing pilar cyst. Histologically, this tumor may mimic squamous cell carcinoma (SCC). The standard of treatment is surgical excision with a margin of normal tissue. As most of the patients are not good surgical candidates, Radiotherapy plays an important role in the management of this tumor. We report a case of PTT on the occipital region of a 79-year-old female with its brief overview. In view of large size of the lesion and advanced age of the patient, Neoadjuvant Radiotherapy was planned and delivered. A near complete response of the lesion to radiotherapy was observed with good cosmesis. We discuss the role of radiotherapy in the management of PTT in various settings with literature review.
Keywords: *Proliferating Trichilemmal Tumor, Proliferating Pilar Tumor, Radical Radiotherapy, Neoadjuvant radiotherapy, outer root sheath tumor, skin cancer, scalp tumor, cosmesis.*

Introduction

Proliferating Trichilemmal Tumor (PTT), is a rare, benign neoplasm arising from the outer root sheath of the hair follicle. It usually arises from pre-existing pilar cyst. It is characterized by small, solitary, asymptomatic lesion which increases in size rapidly. It tends to occur in the fourth to eighth decades of life, and has a predilection for women. The typical histological features are trichilemmal keratinization and lack of granular layer [1, 2]. It occurs most commonly on the scalp in 90% of the cases. The rate of local recurrence and regional lymph node metastasis has been estimated to be between 3.7 - 6.6% and 1.2 - 2.6%, respectively [3].

On the contrary, Malignant Proliferating Trichilemmal Tumors (MPTT) exhibit cellular dysplasia and invasion into the surrounding connective tissue. Histologically they have high mitotic counts, cytological and architectural atypia, cellular pleomorphism, necrosis, infiltrating margins and aneuploidy. The rate of metastasis is as high as 25%[3].

The mainstay of treatment of PTTs include total excision with adequate margins. Radiotherapy can be used in Neoadjuvant, Adjuvant, Definitive as well as in Palliative setting. Hence we report this rare case of PTT with unpredictable biological behaviour, which is frequently confused with SCC with respect to the role of Radiotherapy and treatment outcomes.

Case Report

A 79 year old female patient presented with complaints of expanding swelling in occipital region of scalp since 3 years which was insidious in onset and gradually progressive. It was initially of size 3 cm and progressed to size of 14 cm at presentation. It was not associated with pain, ulceration, loss of sensation, discharge or bleeding. On clinical examination, the lesion was approximately 15 x10 x 10 cm, soft to firm in consistency, lobulated, non-ulcerated, non-mobile, no bleeding or discharge present. It extended superiorly till parietal region and inferiorly till root of the neck (Figure 1).

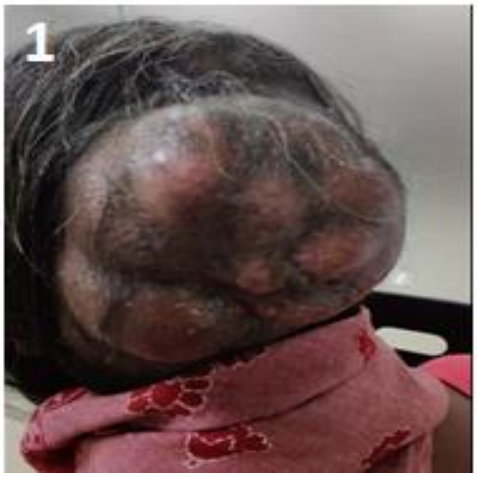

Figure 1: Clincal presentation

Punch biopsy was done which showed lobules of neoplastic epithelium with trichilemmal keratinization evidenced by presence of few peripheral basaloid cells and large keratinocytes with ample eosinophilic cytoplasm that develop abrupt keratinization without granular layer resulting in compact orthokeratotic eosinophilic keratin. At places there is presence of epithelial infoldings with pools of trichilemmal type of keratin (Figure 2 A, B, C). As per these features it was diagnosed as Proliferating Trichilemmal Tumor.

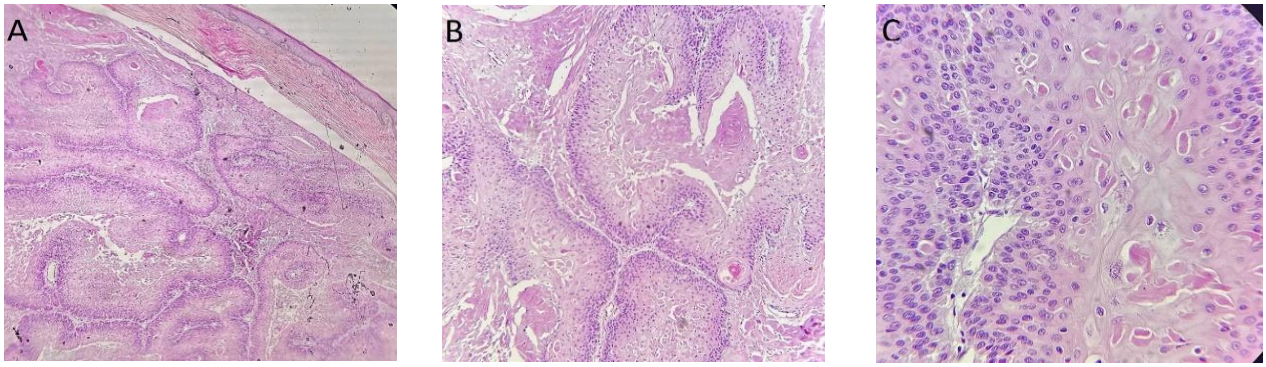

Figure 2(A): Proliferating cystic lesion with trichilemmal differentiation with areas of necrosis (H&E; 4x).
Figure 2(B): Lobules of neoplastic epithelium with trichilemmal keratinization, orthokeratotic eosinophilic keratin (H&E;10x).
Figure 2(C): Trichilemmal differentiation, focal necrosis, monomorphic keratinocytes without significant atypia (H&E; 40x)

CT Brain showed lobular, heterogeneously enhancing solid and cystic space occupying lesion in the right occipital extracranial scalp region of size 14x10x10cm. There were no bone defects except for a focal outer table scalloping (Figure 3, 4).

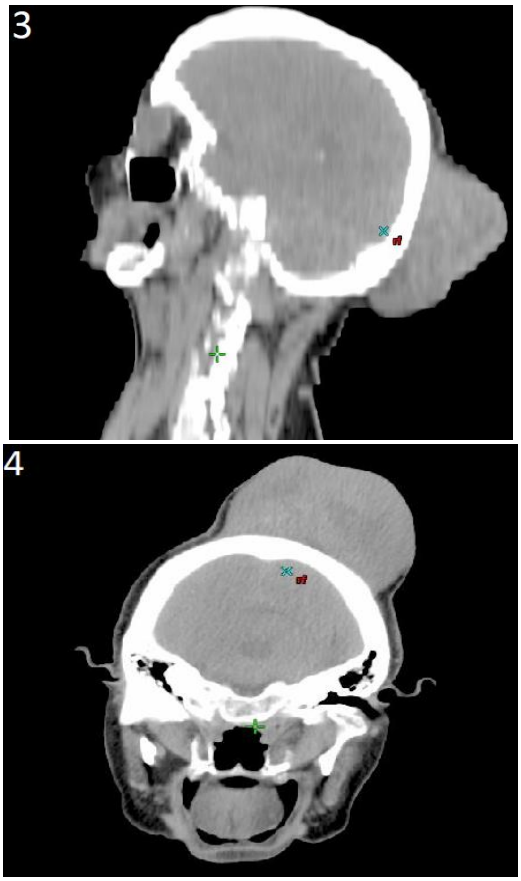

Figure 3,4: Diagnostic CT Images

In view of large size, Neoadjuvant Radiotherapy was initially planned to shrink the tumor prior to definitive surgical excision. She was treated with a total dose of 45 Gy in 15 fractions, 3Gy per fraction (EQD2- 54 Gy for normal tissue, 48.75 Gy for tumor) 5 fractions per week over 3 weeks by 3DCRT technique (Figure 5). Near complete response was seen at six weeks following radiotherapy with excellent cosmesis (Figure 6).

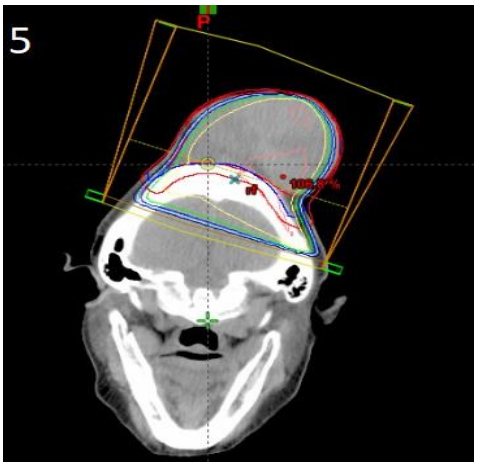

Figure 5: Radiotherapy (3DCRT) planning image

Figure 6: Near complete response at 6 weeks post radiotherapy

Patient defaulted regular follow-up and presented with swelling at same site after 2 years. CT scan demonstrated extra-calvarial, well-defined, lobulated soft tissue attenuating lesion of size 2 x 5 x 3 cm with no cortical bone erosion in the occipital region extending to right of midline-suggestive of recurrent lesion (Figure 7). Wide local excision of the tumor was done. Post op histopathology report confirmed recurrent Proliferating Trichilemmal Tumor with free margins. The patient is on regular follow-up for one year with no recurrence till date (Figure 8)

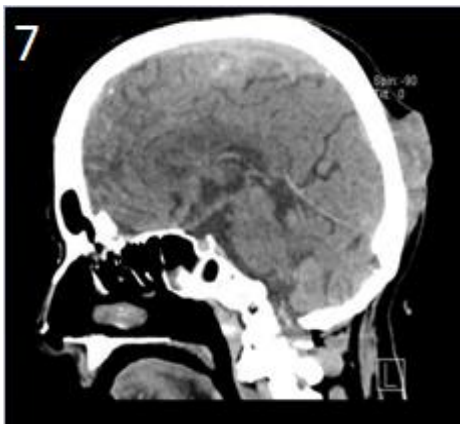

Figure 7: Recurrent lesion at 2 years post radiotherapy

Figure 8: One year post surgery

Discussion

Proliferating Trichilemmal Tumor is a rare neoplasm with less than 100 cases reported in 50 years [4]. It arises from outer root sheath of hair follicle. About 90% of the tumors appear on the scalp in fourth to eighth decade of life and mostly affects women. It is a solitary, asymptomatic, slowly progressing tumor with local invasive nature.

PTT starts with trichilemmal cysts as the original lesion or they may arise de novo. 2% of the trichilemmal cyst can undergo transformation to PTT due to trauma, irritation, or chronic inflammation. CT scan and PET scan can assess the degree of locoregional extension and rule out metastatic involvement. Histologically, this tumor may mimic squamous cell carcinoma (SCC). The distinguishing features that favor PTT over SCC include trichilemmal-type keratinization, the lack of a granular layer of cells, and the absence of a premalignant epidermal lesion. The differential diagnosis includes dermoid cyst, lipoma, hibernoma, pilomatrix carcinoma, SCC etc.

The mainstay of treatment of PTTs include total excision with adequate margins because of the malignant transformation potential and to rule out the possibility of a carcinomatous component that would not be visible in an incisional biopsy [2]. There is currently no clear guideline, since only a few cases have been reported in the literature; However, given the local recurrence rate of 3.7 - 6.6%, other treatment modalities are being investigated, including Mohs surgery and excision with frozen section with margin assessment [7].

To further limit the rate of recurrence, some groups have advocated for adjuvant radiotherapy in the treatment of PTT[2]. The rate of metastasis is as high as 25%[3]. Palliative radiotherapy has been used in metastatic setting. For large size of the lesion, radiotherapy is used as Neoadjuvant modality to reduce tumor size prior to surgery. For advanced age of the patient, cases with potential for surgical complications, such as facial nerve damage, Radical radiotherapy can be opted for treatment.

Duncan Sutherland published a first case report on MPTT treated with Radical Radiotherapy. Complete clinical response was achieved which eliminated the need for aggressive surgery. They concluded that Neoadjuvant radiotherapy had the potential to be an excellent add-on for surgical cases[8].

Cemal Alper Kemaloğlu et. al., published an article of 4 case reports on MPTT of the scalp. In all patients, the tumor developed denovo with local invasion due to late presentation and the treatment became complicated. Adjuvant radiotherapy was recommended for three patients with local invasion, Ki-67>10% and recurrence could be prevented in these patients. One patient with a tumor size of 2 cm with delay in follow-up, presented with systemic metastases without local recurrence. This suggested that the tumor may be

of an aggressive nature independent of the diameter & adjuvant radiotherapy should be applied to all patients.[8]

Our patient first received Neoadjuvant radiation therapy in view of large tumor size with excellent cosmesis post treatment. Patient was not on regular follow-up and presented with swelling at same site after 2 years. She underwent surgery and is on regular follow-up for one year with no complaints.

Conclusion

This case report describes the presentation and course of treatment for Proliferating Trichilemmal Tumor with Neoadjuvant Radiotherapy. Neoadjuvant radiotherapy has the potential to be an excellent modality before surgery. It may also be a good non-surgical alternative in frail patients, elderly patients, large tumors, for cosmesis and to maintain function in certain areas. However a large case series with long-term follow-up is required to understand the complexity of the tumor behaviour and to formulate a guideline to approach these rare tumors.

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