Adjuvant radiotherapy in management of trichilemmal carcinoma of left nasal alae with positive surgical margins: A case report

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Abstract

Background: Trichilemmal carcinoma (TLC) is a rare adnexal malignant tumor developing from the outer root sheath of hair follicles with no distinct clinical features which might clinically be misdiagnosed as actinic keratosis, nodular melanoma, basal or squamous cell carcinoma. Albeit no consensus exists on standard treatment of TLC, tumor excision with adequate clear margins is the current widely accepted treatment consideration with no previous literature on use of radiotherapy (RT) in definitive or postoperative settings.

Case presentation: A 60-year-old woman who was repeatedly treated with cryotherapy located on her left nasal alae for an initial diagnosis of actinic keratosis and diagnosed as TLC at last surgery was referred for RT of microscopic surgical margins. The patient was treated with 6 MeV electron beam RT prescribed to surgical bed plus 1-cm margins at all directions, namely the planning target volume. The total and per fraction doses were 60 and 2 Gy, respectively, which is commonly practiced for any skin tumor with positive margins. The treatment was well tolerated with no acute or chronic complications. The patient was alive with no local, regional, or distant recurrences at the 49 months of her follow-up.

Conclusions: Although the follow-up period is relatively short and further evidence is needed to confirm the exact role of RT in adjuvant treatment of TLC, the outcomes of present rare case of a nasal TLC suggests that adjuvant RT in patients with positive surgical margins may provide satisfactory local tumor control.

Key Words: Trichilemmal carcinoma, Recurrent tumor, Positive surgical margins, Radiotherapy

1. Introduction

Trichilemmal carcinoma (TLC) is a rare adnexal malignant tumor developing from the outer root sheath of hair follicles.[1] Although it may appear as multiple lesions on non-sun-exposed skin, the usual presentation is a non-descript solitary papule/nodule appearing on the sun-exposed, hair-bearing areas, such as the face or ears in elderly individuals.[2] In absence of distinct clinical features TLC may be misdiagnosed as actinic keratosis, nodular melanoma, basal cell carcinoma (BCC), or squamous cell carcinoma (SCC).[3,4]

Wide excision with clear margins is the current standard care option for TLC. Notwithstanding of its malignant appearance on cytological examination, clinically the TLC exhibits an
indolent tumor behavior with rare local relapses after surgical removal of the primary lesion.\cite{2} However, surgery can either be rejected by the patient or be awkward with resultant cosmetic problems on particular locations such as the facial area. Hence, other treatment modalities, including the radiotherapy (RT) may prove beneficial for this patients group.

We herein present an extremely rare case of nasal TLC in a 60-year old lady with the history of repeat cryotherapy for the diagnosis of actinic keratosis who was managed with adjuvant RT for microscopically positive surgical margins after inadequate surgery, as she declined the recommended re-excision procedure.

Figure 1. Histopathologic examination demonstrating; (A) the tumor originating from epidermis with infiltration into the dermis (H&E × 40), (B) present clear- (arrow) and squamous cells (arrow head) (H&E × 100), (C) Epithelial and clear cells (arrow) with significant atypia (H&E × 200), and (D) cells with features of high mitotic activity (arrow) (H&E × 400).

2. CASE PRESENTATION
A 60-year-old woman was first evaluated in dermatology clinics for the complaint of a non-healing, irregularly shaped, grayish papule of 1.5 cm × 1.2 cm on her left nasal alae in December 2004. She was diagnosed as actinic keratosis without necessity for pathologic examination, and the lesion disappeared following one course of cryotherapy. After a disease free interval of 5 years, she was re-admitted to the same dermatology clinics with a similar lesion of 2.0 cm × 1.6 cm and received a second course of cryotherapy in October 2009. But unfortunately, a second recurrence at the same location was experienced by the patient in August 2011 which was managed again with third course of cryotherapy with no response at first month of follow-up. Than she was assessed by a plastic surgeon and tumor was removed surgically in September 2011. On gross examination a tumor mass of 1.5 cm × 1.3 cm × 0.8 cm with histopathologically tumor positive margins was reported. Histopathologic examination demonstrated the tumor that originates from epidermis and infiltrates the dermis with the histological features of actinic dermal degeneration and accompanying intense lymphocytic infiltration, hyper- and para-keratosis, achantolytic cells, atypical basaloid cells expanding through dermis (see Figure 1a). Formation of peripheral palisades with squamous cells and malignant clear cells (see Figure 1b) with significant cellular atypia (see Figure 1c) and frequent mitotic
activity (see Figure 1d) were accounted to be evident (see Figure 1). Based on these features her final diagnosis was TLC. Therefore, the patient was recommended to undergo re- 
operation for wide excision to attain adequate clear margins, 
which was declined by the patient due to cosmetic concerns.

After her referral to our clinic for RT, staging workup with 
neck, chest, and abdominal computerized tomography scans uncovered no regional or distant metastasis. Than between 
the October and November 2011, the patient was treated 
with 6 MeV electron beam RT prescribed to surgical bed 
plus 1-cm margins at all directions, namely the planning 
target volume. The total and per fraction doses were 60 and 
2 Gy, respectively, which is commonly practiced for any skin 
tumor with microscopic positivity. The treatment was well 
tolerated with no acute or chronic complications. The patient 
was alive with no local, regional, or distant recurrences at the 
49 months of her follow-up.

3. DISCUSSION

The term of TLC was first proposed for a histologically 
invasive, cytologically atypical clear cell neoplasm of ad-
nexal keratinocytes which is in continuity with the epider-
mis and/or follicular epithelium by Headington in 1976.[11] 
With a slight male predilection,[2] the TLC which is a rare 
adnexal malignant tumor developing from the outer root 
sheath of hair follicles, frequently occurs on sun-exposed 
skin of elderly individuals; face and ears being the common-
est sites.[1, 2] Actinic damage, burn scar formation, long term 
low dose irradiation exposure and malignant transformation 
from trichilemmoma have been hypothesized,[15–81] but the 
exact pathogenesis of TLC still remains vague. Although it 
is difficult to remark on the exact causative, particular for the 
case presented here, vicinity of actinic dermal degeneration 
on histopathologic examination suggest the solar damage as the 
highly probable underlying cause.

Histopathologically, TLC is portrayed by presence of rel-
atively well circumscribed multiple intradermal lobules or 
trabeculae with peripheral palisading basaloïd cells in con-
gruity with the epidermis. Lobular proliferation centered on pilosebaceous structures composed of polygonal clear cells 
with abundant clear, glycogen rich cytoplasm and prominent 
nucleoli are typical.[11] Presence of high mitotic index and 
striking cytological atypia may grant the impression of a 
high grade, conceivably aggressive malignancy. In our case, 
besides the typical histopathological appearance on the mi-
oscopic examination presence of both cellular atypia and 
high mitotic index were the factors landing support for TLC 
diagnosis.

The uncommon TLC lacks characteristic clinical features 
on inspection and typically presents as a solitary grayish to reddish brown or flesh colored ulcerative nodule.[2] In 
this manner, patients may be mistaken for either benign or 
malignant cutaneous tumors including the actinic keratosis, 
nodular melanoma, BCC, or variants of SCC. Moreover, 
similar with the present case, patients may be inappropri-
ately managed due to this misdiagnosis and may experience 
sobering recurrences. As being what is indicated, the rarely 
suspected TLC ought to be remembered in differential dia-
gnosis of any cutaneous lesion and biopsy should strongly 
be recommended for accurate diagnosis and guidance of the 
subsequent interventions.

The TLC is a malignant but a relatively indolent tumor with 
exceedingly rare local failures and distant metastasis after 
surgery.[2] Albeit no consensus exists on its standard treat-
ment, tumor excision with adequate clear margins is the 
accepted as the current standard consideration. On the other 
hand, surgery may be cosmetically problematic on certain 
locales including the nose, or may simply be declined by the 
patient, impacting the need for alternative noninvasive 
procedures. In this setting, regarding its well-established 
efficacy in BCC and SCC external beam RT appears to be 
an appropriate treatment option for such patients.[9, 10] To 
our best information, this is the first report on successful 
treatment of TLC with RT. We decided to prescribe 50 Gy 
(2 Gy/fr) in view of the way that it is in the commonly 
practiced total dose (45–50 Gy) and fractionation (1.8–2 Gy) 
ranges recommended for any tumor type with microscopi-
cally positive margin(s). Albeit additional proof is justified 
to conclude firmly, considering the 49 months of recurrence 
free interim subsequent to the use of RT, it is reasonable to 
assume that the radiosensitivity of TLC is similar with other 
skin tumors, at least at the microscopic disease setting.

4. CONCLUSIONS

This is the first instance of rarely reported nasal TLC which 
was treated successfully with external beam RT for positive 
surgical margins. The present case is moreover important 
for impacting the need of biopsy for accurate diagnosis of 
TLC, and hence for its appropriate treatment, in absence of 
characteristic clinical features.

CONFLICTS OF INTEREST DISCLOSURE

The authors declare no conflicts of interest.
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