CASE REPORT

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 nondescript 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

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indolent tumor behavior with rare local relapses after surgical removal of the primary lesion.[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.

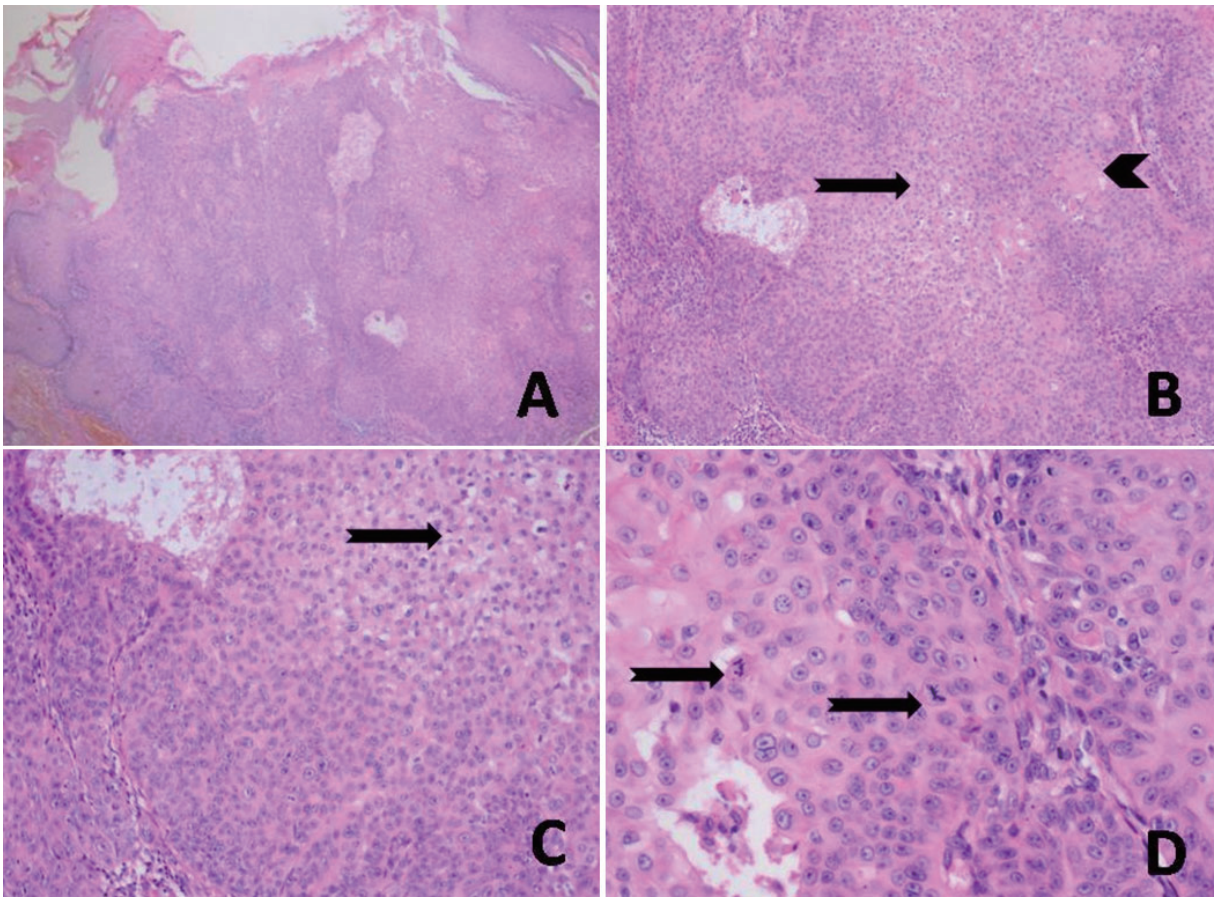

Figure 1. Histopathologic examination demonstrating; (A) the tumor originating from epidermis with infiltration into the dermis (H&E × 40), (B) present clear- (arrow) and squamous cells (arrow head) (H&E × 100), (C) Epithelial and clear cells (arrow) with significant atypia (H&E × 200), and (D) cells 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 lymphoctic 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 adnexal keratinocytes which is in continuity with the epidermis and/or follicular epithelium by Headington in 1976.[1] 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 commonest sites.[1,2] Actinic damage, burn scar formation, long term low dose irradiation exposure and malignant transformation from trichilemmoma have been hypothesized,[5–8] 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 relatively well circumscribed multiple intradermal lobules or trabeculae with peripheral palisading basaloid cells in congruity 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.[1] 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 microscopic 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 inappropriately 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 diagnosis 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 treatment, 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 microscopically 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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