

BRIEF ARTICLE

Rare Mucosal Lip Atypical Fibroxanthoma Treated with Mohs Micrographic Surgery

Tara Howard, BS¹, Taraneh Matin, DO², John Howard, DO³, Weiss Eduardo, MD⁴

¹ Nova Southeastern University-KPCOM, Fort Lauderdale, FL

² Larkin Community Hospital - Palm Springs Campus, Dermatology, Hialeah, FL

³ Hollywood Dermatology & Cosmetics Specialists, Hollywood, FL

⁴ Hollywood Dermatology & Cosmetics Specialists, Hollywood, FL

ABSTRACT

Atypical fibroxanthoma (AFX) is a rare dermal neoplasm of low-intermediate malignant potential found almost exclusively in the non-mucosal regions of the head and neck in light-skinned elderly males who have a history of significant sun exposure. Due to its risk of misdiagnosis of more common skin lesions and possibility of metastases, AFX requires resection with either Mohs Micrographic Surgery (MMS) or wide local excision (WLE). The purpose of this brief article is to discuss the best comprehensive treatment for a lower lip AFX using MMS versus WLE.

INTRODUCTION

Atypical fibroxanthoma (AFX) is a rare (0.002% of all non-melanoma skin cancers) cutaneous neoplasm of fibrohistiocytic mesenchymal origin and low-intermediate malignant potential. Recurrence and local invasion may occur; however, the clinical outlook is excellent as metastatic potential is rare (0.5%). AFX presents as a rapidly growing, solitary, red-pink, firm nodule with a mean diameter less than 2 cm and occasionally may ulcerate or bleed. These tumors present almost exclusively in the non-mucosal regions of the head and neck in light-skinned elderly males who have a history of significant sun exposure.

Ultraviolet (UV) radiation exposure leading to p53 mutations and cyclobutene pyrimidine dimers is the primary risk factor for development of AFX. These tumors possess a predilection for sun-exposed areas in

individuals with actinically damaged skin or previously treated skin cancer. Further evidence supporting UV-induced pathogenesis is the higher incidence of AFX in patients with xeroderma pigmentosum (XP), a disease characterized by mutated DNA repair mechanisms. Other, less common risk factors include prior radiation, burns, trauma or immunosuppression.

We report a 73-year-old female with a lower lip mucosal AFX. A literature search of lower lip AFX revealed only one case of malignant AFX on the lower lip of an elderly male and one on the lower lip in a male pediatric patient with a history of XP. The location of AFX on the lower mucosal lip makes this case notably rare.

CASE REPORT

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A 73-year-old female with a history of significant sun damage and non-melanocytic skin cancer presented to the clinic with the complaint of an enlarging lower lip lesion that did not improve with the use of OTC products. Examination revealed a 1.6 x 1.6 cm pink indurated nodule on the right medial, inferior vermillion of the labial mucosa (Figure 1). The patient underwent a shave biopsy which revealed an atypical spindle cell tumor positive for CD10 and SMA. Desmin, S100, CK-AEa/AE3, Ber-EP4 and P63 were negative. A final diagnosis of AFX was made.



Figure 1.

The patient was treated in 2 stages of Mohs Micrographic Surgery [MMS] with a surgical margin of 0.2 cm. Postoperative dimensions measured approximately 2 x 2 cm (Figure 2). A mucosal advancement flap was used to repair the defect (Figures 3). At one-week follow-up the surgical site revealed a well-healed, linear scar absent of drainage or dehiscence and sutures were removed (Figure 4).



Figure 2.



Figure 3.

DISCUSSION

AFX requires aggressive treatment due to its risk of misdiagnosis and possibility of metastases. Patients treated with MMS have a recurrence rate of 2-4.6%, often within 1-3 years of resection. A retrospective study compared 91 AFX patients treated with either MMS or WLE [wide local excision] to determine which modality provided the best comprehensive treatment; of the 59 patients treated with MMS, recurrence did not occur, whereas the 23 patients treated with WLE had a recurrence rate of 8.7%. The median margin for MMS clearance was found to be significantly smaller [0.4 cm] in comparison to 2 cm for a



Figure 4.

WLE clearance of 95%. Therefore, MMS with complete resection is preferred over WLE because the recurrence rate is reduced, the incidence of metastases is lowered and more facial tissue is preserved providing the greatest aesthetic outcome^{1,2}.

To maintain normal anatomy, optimal function, and cosmetic integrity of the inferior vermilion labial mucosa, we considered mucosal advancement flap as the most ideal repair technique. Our patient responded well to both MMS and mucosal advancement flap repair; not only was her neoplasm effectively removed and treated, the choice in repair provided for a smooth recovery and superior aesthetic results compared to alternative techniques.

CONCLUSION

Isolated AFX on the lower lip mucosa is exceedingly rare as there has been only one case report in 1975. Patients with a history of AFX should undergo bi-annual skin examinations for signs of recurrence or presence of new skin malignancies, as these patients represent a high-risk group

for development of UV associated malignancies.

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Corresponding Author:

Howard, Tara, BS
3850 Hollywood Blvd #301
Hollywood, FL 33021
Email: th1302@mynsu.nova.edu

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