

# The various treatment modalities of dermatofibrosarcoma protuberans

Aatman Shah<sup>1</sup> MD, Michael Tassavor<sup>1</sup> MD, Sayesha Sharma<sup>2</sup> BS, Richard Torbeck<sup>1</sup> MD

Affiliations: <sup>1</sup>Department of Dermatology, Mount Sinai School of Medicine, New York, New York, USA, <sup>2</sup>St. George's University School of Medicine, True Blue, Grenada

Corresponding Author: Richard Torbeck MD, 1468 Madison Avenue, New York, NY 10029, Tel: 212-241-6500, Email: [Richard.Torbeck@m Mountsinai.org](mailto:Richard.Torbeck@m Mountsinai.org)

*Keywords: chemotherapy, dermatofibrosarcoma protuberans, Mohs, radiation, surgery*

To the Editor:

We have read, with great interest, an article published in the *Dermatology Online Journal* by Patel et al. entitled "Report of rare case of dermatofibrosarcoma protuberans in the buccal mucosa: review of diagnostic, histopathological, and immunohistochemical criteria" [1]. In this paper, the authors report on a rare case of dermatofibrosarcoma protuberans (DFSP) found within the oral mucosa of a patient. The authors also do an excellent job of reviewing the diagnostic, histopathological, and immunohistochemical criteria for DFSP. In addition to understanding diagnostic nuance, it is also important for physicians to be aware of the various treatment modalities for DFSP to educate patients in the decision-making process. This letter is written to briefly discuss the various potential treatment options for DFSP.

The primary treatment modality is surgical, and data has demonstrated that Mohs Micrographic Surgery (MMS) is preferred over wide local excision (WLE) owing to the unpredictable subclinical extension of DFSP. In a cohort of 79 patients treated with either MMS or WLE, Paradisi et al. demonstrated that there was a 0% recurrence rate for those treated with MMS versus a 13.2% recurrence rate for those treated with WLE [2]. Furthermore, pooling their data with all available cases in the literature revealed only 6 recurrences out of 463 cases treated with MMS (1.3%, 95% CI 0.5-2.8%) versus 288 recurrences out of 1,394 cases treated with WLE (20.7%, 95% CI 18.6-22.9%) [2]. The relative risk of recurrence for WLE versus

MMS patients was 15.9 (95% CI 7.2-35.5) [2]. Nevertheless, it is important for readers to know that WLE may still be performed when MMS is not available or in select situations. Wide local excision may be performed with 3cm margins. However, local recurrence can occur even with clear surgical margins. Modified WLE with horizontal sectioning as opposed to vertical processing ("bread-loafing") can be performed to decrease the likelihood of local recurrence.

Aside from surgical approaches, it is important for clinicians to be aware of adjunctive treatment modalities. The chemotherapy agent imatinib mesylate, an oral tyrosine kinase inhibitor, is used for recurrent, unresectable, and metastatic DFSP rather than for curative intent [3]. Imatinib mesylate competitively inhibits ATP binding to the PDGF-beta receptor, which slows down kinase activity, limits growth of the tumor, and promotes apoptosis. Patients with the t(17;22) translocation demonstrate a greater response to imatinib mesylate with studies suggesting a response rate of about 65% [3]. Screening for this translocation can be performed using fluorescent in situ hybridization or reverse transcription-polymerase chain reaction. For cases in which imatinib is unsuccessful, there have been a few reports in which patients responded to sunitinib [4], an inhibitor of multi-tyrosine protein kinases, or sorafenib [5], a BRAF and vascular endothelial growth factor (VEGF) receptor inhibitor.

Aside from chemotherapy, radiation therapy may also be efficacious for unresectable or recurrent tumors and adjuvant radiation may decrease the risk of local recurrence. Du et al. published a retrospective analysis of 184 patients with DFSP and

demonstrated in their matched cohort that 5 year disease-free survival was significantly higher in the surgery and radiation group than in the surgery group alone (88.1% versus 56.2%,  $P = 0.044$ ) [6]. Haas et al. further demonstrated that 21 patients who were treated with WLE had a local control of 67% whereas 17 patients who were treated with surgery and radiation had a local control of 82% [7]. In summation, the available data has shown that postoperative adjuvant radiation therapy may reduce the risk of recurrence when clear surgical margins are not obtained.

The diagnosis and management of dermatofibrosarcoma protuberans requires a

multidisciplinary approach to treatment consisting of a surgeon, oncologist, radiologist, and physical therapist. We hope that this letter will help providers discuss the risks and benefits associated with the various treatment options with their patients. Although MMS appears to have superior outcomes in the treatment of DFSP, awareness of all treatment modalities will aid in clinical decision-making and thus promote better healthcare for our patients.

### Potential conflicts of interest

The authors declare no conflicts of interest

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