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Orofacial Granulomatosis Treated with Low-dose Radiation Therapy: A Case Report

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Abstract We are reporting a rare chronic irritating, incapacitating condition that was controlled by low-dose radiation treatment in the case here presented. A 58 year-old woman had a 2½-year history of painful orofacial granulomatosis was referred to our practice after many different treatment options failed control the disease. Presenting severe difficulty of swallowing, discomfort, labial swelling and oral ulcerations it was decided to give a trial of low-dose radiation treatment. Excellent treatment outcome was observed with significant diminution in swelling, ulceration, thickening and induration. Nine months after treatment, the patient is back to her normal self and maintains a high quality of living. Low-level laser therapy may be an alternative option, however not conclusive. The experience of low-dose radiation treatment of orofacial granulomatosis has not been reported clearly in the literature, but this case may indicate it as. Therefore we would like to share this information, suggesting that low-dose radiation treatment may be an alternative treatment for orofacial granulomatosis, and that should be extensively explored both in research and clinical trials when all other options of treatment have failed.

Kewords Orofacial Granulomatosis, Radiation Therapy

1. Introduction

Orofacial granulomatosis encompasses different granulomatous conditions in the oral cavity. OFG presents with labial swelling, oral ulcerations, gingival enlargement, and cobblestoning mucosa. This condition was first described by Wisenfeld in 1985 (1). The etiology of this condition is unknown, and at a minimum requires the exclusion of sarcoidosis. The typical histopathologic features include noncaseating giant cells, epithelioid granulomas and lymphedemas. Different therapeutic approaches have been reported including topical and systemic corticosteroids, intralesional injection of triamcinolone, monoclonal antibodies, and chemotherapeutic agents including thalidomide, methotrexate, topical tacrolimus and clofazimine. Surgical treatment with cheiloplasty is not recommended because of the high rate of recurrence (2-8). Low level laser therapy (9) is an alternative therapeutic and noninvasive treatment particularly for the treatment of ulcerative lesions which are not responding to conventional corticosteroid therapy, but such therapy has not been the subject of extensive research neither greatly reported at this time. Hereby we would like to report a case that was treated with low-dose radiation treatment with considerable success.

1.1. Case Report

A 58 year-old lady was seen on 5/23/12 with a 2½ -year history of painful swelling of her lips, left buccal mucosa and left side of the tongue (Figure 1). The patient additionally has a history of rosacea affecting both cheeks. Biopsies revealed dense plasma cell infiltration, with clusters of histiocyte-forming granulomatosis, however no definitive pathologic diagnosis was made. Special stains for organisms were negative, and immunohistochemistry for kappa and lamda light chains showed no abnormal findings. Glossal biopsy confirmed ulceration with associated granulation tissue. After considerable evaluation and discussion, the clinical diagnosis of OFG was made. The patient was treated with prednisone 40 mg daily with some mild improvement, but the condition progressively worsened over a span of 2½ years. The patient was also treated with Remicade and Valtrex E, with minimal improvement. Due to trismus resulting from persistent and considerable swelling and induration of her lips, buccal mucosa, tongue and floor of the mouth, she was unable to maintain adequate oral intake; only able to open her mouth about 2.0 cm. After careful consideration the patient was recommended for a trial of low-dose radiation treatment, at which time she was referred to our practice. Radiation was given in May through June of 2012. She received a daily dose of 150 cGy and a total of 2800 cGy. Treatment was completed on 6/27/12. The patient

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tolerated treatment well, with gradual improvement. When seen on follow-up examination in 9/2012 there was excellent response, with significant diminution in swelling, ulceration, thickening and induration (Figure 2, three months later. Figure 3, nine months later). The patient was able to open her mouth much wider and was able to swallow food and drink liquids. Her clinical response was considerable with this low-dose radiation treatment.

2. Discussion

Different types of OFG have been described including Melkerrson-Rosenthal syndrome which includes orofacial edema, facial palsy and fissured tongue. Spontaneous remission of OFG is extremely rare and treatment may require a special diet with exclusion of food, avoidance of cosmetics containing possible allergens, or removal of suspected dental material allergens such as dental amalgam. Immunosuppressive therapies have been proposed, but are associated with severe side effects and questionable efficacy. Anti TNF-alpha therapy has been used with some degree of success in conjunction with both thalidomide and infliximab. The use of radiation therapy has not been documented well in the literature, and at the time this case report was written no definitive retrospective or prospective studies have been reported. Our trial of low-dose radiation treatment in this patient was out of desperation, as her disease was progressing over a span of 2 ½ years with rapid deterioration of her clinical condition despite the latest medical therapy. The treatment was tolerated reasonably well, without significant mucosal reaction. Three months following completion of therapy, there was dramatic reduction in swelling of the lips and extent of mucosal and glossal induration. It is believed that this is the first documented case of OFG treated with low-dose radiation, demonstrating considerable response. The patient will be followed more carefully for the long-term sequelae of this low dose radiation treatment.

Figure 1. Prior to radiation treatment showing swelling, indurations and erythema around orofacial area

Figure 2. Three months after radiation treatment with significant improvement

Figure 3. Nine months after treatment, returned to her normal orofacial appearance

3. Conclusions

Orofacial granulomatosis is a rare and difficult condition to manage. The current treatments options are limited, and sometimes of questionable efficacy. In an advanced case such as ours, the patient may benefit from low-dose radiation treatment, without any significant side effects. Further exploration and clarification of the role of low-dose radiation in the treatment of debilitating orofacial granulomatosis is warranted, based on the efficacy demonstrated in this case.

Abbreviations: OFG= orofacial granulomatosis.

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