Factitial pemphigus-like lesions

Ali Taghavi Zonuz [1], Nathaniel Treister [2], Fahimeh Mehdipour [1], Ramin Mostofizadeh Farahani [3], Richard Shane Tubbs [4], Mohammadali Mohajel Shoja [5]

(1) Assistant Professor, Department of Oral Medicine, Faculty of Dentistry, Tabriz University of Medical Sciences, Tabriz, Iran
(2) Associate Surgeon, Division of Oral Medicine and Dentistry, Brigham and Women's Hospital, and Instructor, Division of Oral Medicine, Infection, and Immunity, Harvard School of Dental Medicine, Boston, MA, USA
(3) Research Assistant, Faculty of Dentistry, Tabriz University of Medical Sciences, Tabriz, Iran
(4) Assistant Professor, Department of Cell Biology, University of Alabama at Birmingham, USA
(5) Research Assistant, Faculty of Medicine, Department of Anatomy, Tabriz University of Medical Sciences, Tabriz, Iran

Correspondence:
Dr. Ramin Mostofizadeh Farahani
School of Dentistry, Tabriz University of Medical Sciences,
Golgasht St., Daneshgah St., Tabriz, Iran.
E-mail:r.mostofi@gmail.com

Received: 1-07-2006
Accepted: 17-12-2006

Zonuz AT, Treister N, Mehdipour F, Farahani RM, Tubbs RS, Shoja MM. Factitial pemphigus-like lesions. Med Oral Patol Oral Cir Bucal 2007;12:E205-8.

Indexed in:
-Index Medicus / MEDLINE / PubMed
-EMBASE, Excerpta Medica
-SCOPUS
-Indice Médico Español
-IBECS

ABSTRACT

The maxillofacial region is rarely subjected to self-inflicted conditions such as factitious disease. Nasal ulceration, facial emphysema, periorbital ecchymosis, mandibular subluxation, gingival and mucosal ulceration, dental and salivary gland pain and glossopharyngeal neuralgia have been reported as possible manifestations of factitious disease. We report a case of a young woman who presented with unilateral bullous and ulcerative oral and erythematous facial lesions that were initially diagnosed as pemphigus vulgaris but was later determined to be secondary to self-inflicted injuries. To the best of the authors' knowledge, this clinical scenario has not been previously reported in the context of a factitious disease and, therefore, may be considered in the differential diagnosis of oral vesiculobullous disorders.

Key words: *Bullous, erythematous, facial, oral, factitious disease, pemphigus vulgaris.*

INTRODUCTION

Factitious disorders are characterized by intentionally produced or feigned psychological or physical signs and/or symptoms in order to assume a sick role in the absence of any known external incentives for the behavior (1). The so-called Munchausen syndrome is characterized by patients who chronically seek medical attention and present with predominately physical findings, however, both terms are often used interchangeably (2). The maxillofacial region is rarely subjected to these self-inflicted injuries. We describe a young woman with factitious disorder who presented with unilateral bullous and ulcerative oral and erythematous facial lesions that was initially misdiagnosed as pemphigus vulgaris.

CASE REPORT

A 17-year-old female presented to the oral medicine department with a complaint of "sore gums" . Her past medical history was unremarkable. Physical examination revealed a young lady who was in no acute distress. She presented with erythema of the left facial skin as well as unilateral bullous and ulcerative lesions of the oral mucosa on the ipsilateral side (Fig. 1a,b). The facial erythematous macular lesions were well-demarcated and covered the entire left cheek with no associated crusting, ulceration, or swelling. The oral lesions, characterized by desquamation, erythema and ulceration, involved the facial aspects of the maxillary and mandibular gingiva and alveolar mucosa from approximately the right lateral incisor to the left second molar. Her oral hygiene appeared adequate. The rest of the examination was otherwise unremarkable. The patient had no other areas of erythema or ulceration.

Upon questioning, she denied the use of any harmful materials or overzealous toothbrushing all of which were considered as possible etiological factors. When the patient was asked about the onset of her condition, she had no definite answer. Laboratory findings with reference to hematological and blood chemistry tests were all within normal limits.

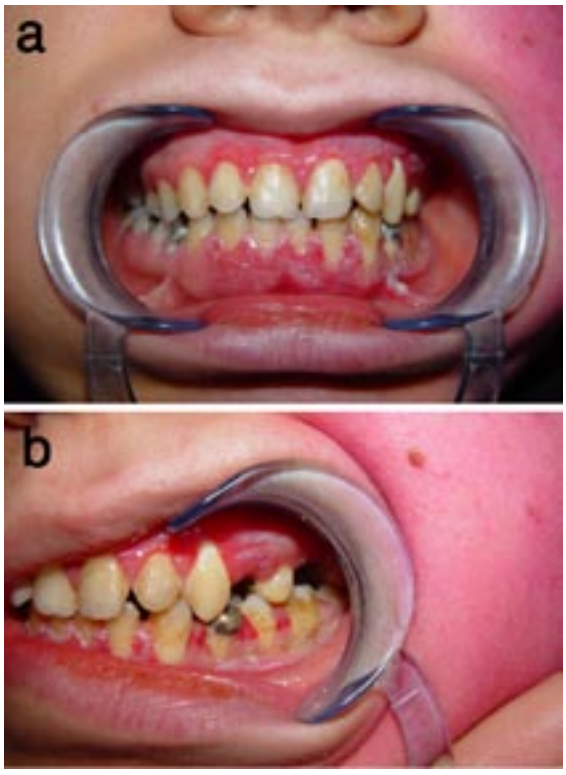

Fig. 1. The anterior (a) and lateral (b) views of the orofacial lesions. Unilateral ulcerative oral lesions and the erythematous facial lesion are evident.

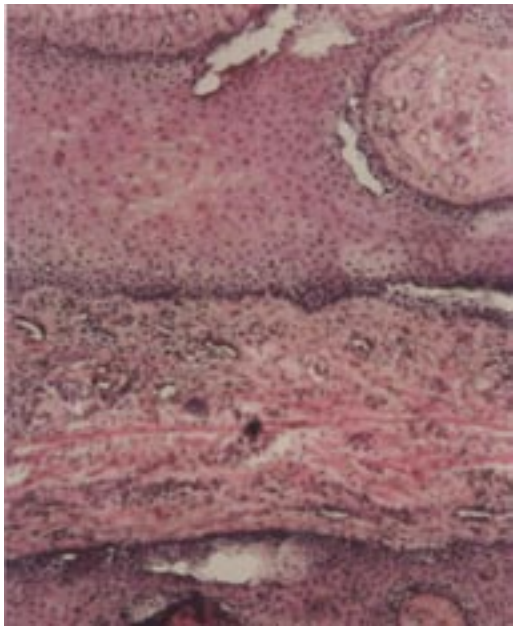

Fig. 2. Photomicrograph demonstrating intra-epithelial blisters as well as inflammatory infiltration throughout the connective tissue (H&E, magnification x20).

A punch biopsy specimen was obtained from the affected oral mucosa adjacent to the mandibular second premolar containing both normal and lesional tissue. The light microscopic examination of the specimen showed intra-epithelial blister formation as well as sub-epithelial inflammatory infiltration suggestive of pemphigus vulgaris (Figure 2). A second specimen was sent for direct immunofluorescence examination. With a presumptive diagnosis of pemphigus vulgaris, 80 mg per day of prednisone was initiated.

At follow-up three days later, the facial lesion was noted to have increased considerably in size and color intensity. These changes in the lesion characteristics were unusual and unexpected. After application with alcohol-impregnated gauze, the erythema diminished considerably. Following further counseling, the patient presented us with a bottle of carpet dye from her purse. Application of the dye to the normal skin produced a color closely resembling that of the facial lesion suggesting a feigned scenario. The oral lesions were essentially unchanged. The steroid therapy was discontinued and the patient and her family were then asked to consult with a psychiatric specialist. The pathology laboratory was notified to disregard the sample sent for direct immunofluorescence.

On psychological interview, the patient explained that she is the youngest of four children in an educated family. She had two older brothers who were engineers and an older sister who was a pharmacologist. She mentioned that university education was highly valued in her family and that her parents expected her to be accepted in a prestigious major at the university. The uncertainty regarding her ability to pass the university entrance exam had been a major source of stress in her life during the last several months. The diagnosis of factitious disorder was made after the patient confirmed the use of carpet dye and also the self-inflicted nature of her facial/oral lesions. Thereafter, the psychological consultation was continued. Two months later the patient was contacted and her sister confirmed the resolution of the orofacial lesions.

DISCUSSION

Self injurious behavior (SIB) is a complex disorder. Different theories have been suggested regarding its etiology. Biological causes such as Lesch-Nyhan and Gilles de La Tourett syndromes, autism, familial dysautonomia and mental retardation have been well-recognized (3-7). On the other hand, functional theories maintain that escape or attention seeking through SIB, which may arise in stressful situations, may be etiological factors especially in the absence of any known biological factors. In our case, the chronic social stress associated with her university entrance exam and family expectations might have caused the patient to use the SIB as a means of escape or excuse. More importantly, as has been previously reported, becoming the focus of her family's attention appeared to be a secondary gain (2, 8, 9).

Competition among siblings for parental love has been an important driving force in human evolution. The fact that parents provide their children with a series of niches

or compartmentalized emotional, behavioral and psychological microenvironments has been suggested previously (10, 11). Even when the parents do not favor one child over another, the competition between siblings continues over issues such as cultivation and exploitation of these family positions (12). In the present case, the educational status and the social prestige and privilege gained through educational achievement were significant factors affecting the dynamics of the patient's family.

Stewart and Kernohan classified factitial oral lesions (FOL) as type A being those superimposed upon a pre-existing lesion, type B involving the injuries secondary to an established habit, and type C as those of an unknown or complex etiology (13). The FOL in the present case should be categorized as type C due to the complex psychological etiology and lack of preexisting lesions or habits. Different variants of FOL have been described including factitial gingivitis, factitial ulcer, factitial periodontitis, and auto-extraction (4, 14-16). The etiology of FOL is diverse. Habits such as digit sucking, fingernail and lip biting, bruxism, chemical injury and placement of foreign objects in the mouth may be responsible for this phenomenon (14, 15, 17-24). In our patient, we could not determine the exact cause of oral lesions however the use of an irritant or a caustic material seems probable.

There have been few reports of oral factitial disease being mistaken for more serious medical conditions. Barrett et al. reported a case that was initially mistaken clinically for an oral vesiculobullous disease, however, histopathology, direct immunofluorescence, and serology were all found to be negative (20). In several reported cases clinical and histopathological findings were consistent with mucous membrane pemphigoid, and only with negative direct immunofluorescence and lack of response to systemic steroid therapy was the true diagnosis obtained (8,25). Other reports have described findings highly suspicious for malignancy, with one initially considered to be metastatic liposarcoma and another mistaken histopathologically for either leukemia or lymphoma, only ruled out after multiple tissue biopsies, molecular studies and bone marrow biopsy (26,27). To the best of our knowledge, there have been no previous reports of oral factitious disease presenting with both clinical and histopathological findings consistent with pemphigus vulgaris. These reports stress the importance of obtaining a detailed clinical and social history, careful interpretation of laboratory results, and consideration of factitious disease in the differential diagnosis of cases that either present atypically, have a poor or incomplete history, or that do not respond to appropriate therapy.

The combination of oral and facial lesions in the context of a factitial disorder has not been previously reported. Discrete factitial facial lesions are well-reported and possibly more common than previously thought (9). Ugurlu et al. found that 18 out of 38 patients with factitious dermatitis had facial lesions and these were more common in women (9). However, there was no mention of oral involvement in this study. Rogers et al. performed a retrospective study on patients referred to a pediatric dermatologist revealing thirty-two patients with artifactual skin disease of whom four patients had presented with erythematous lesions which could be removed by wiping with a swab (28). These lesions were due to applied paint or pigment, a scenario closely resembling the one presented in this report. Facial erythema has also been reported secondary to the application of a hot towel, and in one bizarre case, repeated application of menstrual blood (29,30). The case involving menstrual blood also showed interesting parallels to the present report as both cases were young females with a similar geographic region and under a comparable emotional stress where attention of the family members due to their illness was an important psychosocial or cultural component of the presentation.

Once the diagnosis of factitious disease is made, treatment is complex and challenging and requires a multidisciplinary approach. Implicit comments regarding the nature of lesions and asking the patient whether an injury has occurred previously may be helpful although direct confrontation is often ineffective. Psychological or psychiatric consultation is essential to the success of treatment. According to Schaffer et al. these patients often have a borderline personality disorder and a stressful diathesis may be detected (31). Effective management may require a team of clinicians including oral health, medical, and mental health physicians. We recommend that clinicians should consider factitious disorder in the list of differential diagnosis for oral bullous lesions even if it is pathologically suspected.

REFERENCES

1. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders (DSM-IV). Washington, DC: American Psychiatric Association; 1994. p. 471-5.
2. Zohar Y, Avidan G, Shvili Y, Laurian N. Otolaryngologic cases of Munchausen's syndrome. Laryngoscope 1987;97:201-3.
3. Friedlander AH, Cummings JL. Dental treatment of patients with Gilles de la Tourette's syndrome. Oral Surg Oral Med Oral Pathol 1992; 73:299-303.
4. Mass E, Gadoth N. Oro-dental self-mutilation in familial dysautonomia. J Oral Pathol Med 1994;23:273-6.
5. Saemundsson SR, Roberts MW. Oral self-injurious behavior in the developmentally disabled: review and a case. ASDC J Dent Child 1997; 64:205-9, 228.
6. Cusumano FJ, Penna KJ, Panossian G. Prevention of self-mutilation in patients with Lesch-Nyhan syndrome: review of literature. ASDC J Dent Child 2001;68:175-8.
7. Medina AC, Sogbe R, Gomez-Rey AM, Mata M. Factitial oral lesions in an autistic paediatric patient. Int J Paediatr Dent 2003;13:130-7.
8. Heasman PA, MacLeod I, Smith DG. Factitious gingival ulceration: as presenting sign of Munchausen's syndrome? J Periodontol 1994; 65:442-7.
9. Ugurlu S, Bartley GB, Otley CC, Baratz KH. Factitious disease of periocular and facial skin. Am J Ophthalmol 1999;127:196-201.
10. Plomin R, Daniels D. Why are children in the same family so different from one another? Behavioral and Brain Science 1987;10:1-60.
11. Loehlin J. Genes and Environment in Personality Development. Newbury Park, CA: Sage Publications; 1992.
12. Sulloway F. Born to Rebel: Birth Order, Family Dynamics, and Creative Lives. New York: Vintage; 1997.
13. Stewart DJ, Kernohan DC. Self-inflicted gingival injuries. Gingivitis artefacta, factitial gingivitis. Dent Pract Dent Rec 1972;22:418-26.
14. Pattison GL. Self-inflicted gingival injuries: literature review and case report. J Periodontol 1983;54:299-304.

15. Beckett H, Buxey-Softley G, Gilmour AG. Self-inflicted gingival injury. Br Dent J 1995;178:246.
16. Behrendt A, Vahrson K, Wetzel WE. Consequences of serious oral injury associated with the congenital analgia syndrome. ASDC J Dent Child 1997;64:264-6.
17. Baer PN, Archard HO. Factitial disease of the gingiva and buccal mucosa. Report of a case. N Y State Dent J 1974;40:33-6.
18. Groves BJ. Self-inflicted periodontal injury. Br Dent J 1979;147:244-6.
19. Cristobal MC, Aguilar A, Urbina F, Guerra P, Sánchez de la Paz F, García-Pérez A, et al. Self-inflicted tongue ulcer: an unusual form of factitious disorder. J Am Acad Dermatol 1987; 17:339-41.
20. Barrett AP, Buckley DJ. Covert self-mutilation of oral tissues and skin by mechanical and chemical means. Oral Surg Oral Med Oral Pathol 1988;65:685-8.
21. Greene PR. An unusual self-inflicted gingival injury. Br Dent J 1994; 177:23-4.
22. Creath CJ, Steinmetz S, Roebuck R. A case report. Gingival swelling due to a fingernail-biting habit. J Am Dent Assoc 1995;126:1019-21.
23. Sandhu HS, Sharma V, Sidhu GS. Role of psychiatric disorders in self-inflicted periodontal injury: a case report. J Periodontol 1997; 68:1136-9.
24. Saito T, Koga T. Oral self-mutilation. Psychiatr Serv 2000;51:1052.
25. Kotansky K, Goldberg M, Tenenbaum HC, Mock D. Factitious injury of the oral mucosa: a case series. J Periodontol 1995;66:241-5.
26. Fusco MA, Freedman PD, Black SM, Lumerman H. Munchausen's syndrome: report of case. J Am Dent Assoc 1986;112:210-2.
27. Harrison M, Roberts GJ, Morgan PR, Pinkerton R. Oral self-mutilation masquerading as malignancy. J R Soc Med 1998;91:40-2.
28. Rogers M, Fairley M, Santhanam R. Artefactual skin disease in children and adolescents. Australas J Dermatol 2001;42:264-70.
29. Lamey PJ, McNab L, Lewis MA, Gibb R. Orofacial artefactual disease. Oral Surg Oral Med Oral Pathol 1994;77:131-4.
30. Yanik M, San I, Alatas N. A case of factitious disorder involving menstrual blood smeared on the face. Int J Psychiatry Med 2004;34:97-101.
31. Schaffer CB, Carroll J, Abramowitz SI. Self-mutilation and the borderline personality. J Nerv Ment Dis 1982;170:468-73.