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CASE REPORT

Prosthodontic Rehabilitation of an Oral Pemphigus Vulgaris Patient

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ABSTRACT

Pemphigus vulgaris is a rare autoimmune mucocutaneous disease with high mortality if untreated. Dental management is complicated due to involvement of oral mucosa, increased risk of oral disease and difficulty in rendering dental care. A male patient with exacerbated characteristics was rehabilitated with full mouth restorations following systemic treatment of pemphigus vulgaris. Owing to the nature of the disease and discomfort in providing adequate oral hygiene measures, dietary intake and masticatory function, it is imperative to rehabilitate the patient with well-designed prosthesis. Although these diseases are relatively uncommon, clinicians must be sufficiently familiar with clinical manifestations to ensure early diagnosis and treatment.

Keywords: Pemphigus vulgaris, Mucocutaneous disease, Fixed partial prosthesis.

INTRODUCTION

Phemphigus is a serious autoimmune disease characterized by acantholysis and subsequent blistering of the skin and oral mucosa, mediated by autoantibodies directed against intercellular antigens desmoglein-3 and desmoglein-1.[1-3] There are three immunologic forms of pemphigus; pemphigus foliaceus, pemphigus vulgaris (PV) and paraneoplastic pemphigus.[4] PV accounts for 70 of the cases.[5] The disease is rare with potentially fatal consequences and has a reported incidence of 0.1 to 0.5 cases per 100,000 individuals worldwide per year.[6,7] It is seen worldwide with a predilection of Mediterranean people and Ashkenazi Jews.[5] It is slightly predominant in women and primarily manifests in adults during the fifth and sixth decade of life.[7]

Oral mucosa is most often the initial site, and in many cases the only site, of disease penetration.[6] Any site in the mouth may be involved, but the soft palate, buccal, gingival and lower lip mucosa usually predominate.[8,9] The most common clinical presentation of PV is multiple, chronic ulcerations whose color is red, white or a mixture of both.[10] Affected oral tissues are highly friable with a tendency to bleed and sheer when subjected to minor trauma.[11] Gentle lateral pressure applied to an area adjacent to the affected site forms a blister resulting in a positive Nikolsky's sign.[12] Gingival involvement may manifest as desquamative gingivitis.[13] Patients affected by these chronic, painful ulcers often complain of discomfort with eating spiced or sour foods and performing daily oral functions, including routine oral hygiene.[8] The definitive diagnosis of PV cannot be based solely on clinical examination, as several other oral vesiculobullous and ulcerative lesions have a similar appereance such as lichen planus, pemphigoid and erythema multiforme.[14,15] An incisional, perilesional biopsy containing intact epithelium is required for a definitive diagnosis.[10]

Since development of oral lesions may precede skin lesions, it is not common for a patient having this disease to seek care first from a dentist.[16] The article reports a severe case of PV who was rehabilitated prosthodontically with full-mouth fixed partial prosthesis.

CASE REPORT

A 59-year-old male patient referred to Baskent University, Faculty of Dentistry, Department of Prosthodontics with a complaint of swelling and pain on his left mandibular second molar tooth along with oral ulcers. Medical and present complaint histories were taken at the time of the first consultation, and a complete intraoral, extraoral and radiologic examination was performed (Fig. 1). He had no history of any medical illnesses or allergies and he did not smoke. Extraoral

Fig. 1: Panoramic radiograph of the patient at initial examination

examination was negative for visible skin or ocular lesions. There was no lymphadenopathy. He mentioned that he had difficulties in consuming sour and spicy food. Intraoral examination revealed that the oral mucosa was dry. The patient had full-mouth fixed metal-reinforced-porcelain bridges which had been constructed 3 years ago. After clinical and radiological evaluation, due to acute infection, the mandibular left second molar was endodontically treated following removal of the metal-reinforced crown. Blisters and gingival inflammation were noticed on gingiva around the existing restorations and buccal tissue. The Nicolsky sign was positive. All existing restorations were removed and the patient was referred to the dermatology clinic for an examination (Fig. 2). The dermatology clinic requested for a biopsy, therefore, the patient was referred to the Department of Oral and Maxillofacial Surgery. A full-thickness tissue for biopsy was obtained and sent to pathology. There was no skin lesions described at the time of biopsy. Diagnosis of PV was confirmed by histopathological examination and immunofluorescence analysis.

After administration of a 2 month oral, systemic corticosteroid therapy (Deltacortil, Pfizer Inc., Istanbul, Turkey), the symptomatic oral lesions showed considerable regression. The systemic corticosteroid was prescribed by the dermatologist, starting with 100 mg/day, then the dose was constantly decreased up to 20 mg/day and the patient was released, but continued follow-up.

Meanwhile temporary fixed partial restorations were constructed with supragingival margins. With the oral PV under control, oral rehabilitation was initiated and an informed consent form was signed by the patient related to his prosthodontic treatment. Close to the end of the steroid therapy, all teeth were reprepared with supragingival chamfer margins where possible (Fig. 3). Impressions were made using additional silicone impression material (Elite HD+, Zhermack, Italy). Then, metal-reinforced porcelain fixed-partial restorations were

Fig. 2: Intraoral views after removal of the existing restorations

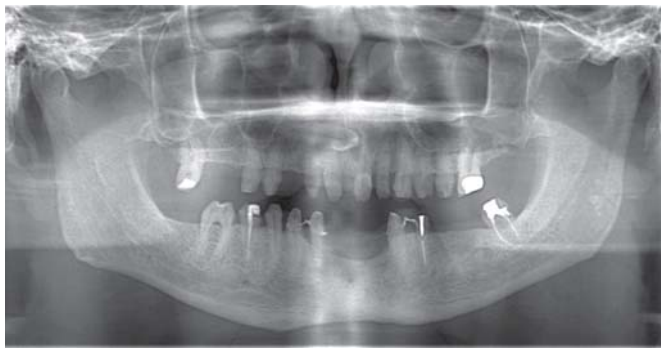

Fig. 3: Panoramic radiograph prior to cementation of the newly constructed restorations

prepared. The new restorations were temporarly cemented (Temporary cement-zinc oxide noneugenol, RelyX Temp NE, 3M ESPE, Germany) for 1 week as a try-out period. Afterward, both the restorations and the teeth were cleaned and cemented permanently using zinc polycarboxylate cement (Adhesor Carbofine, SpofaDental, CZ).

Oral hygiene instructions on effective brushing techniques were demonstrated and utilized for plaque control. A nonalcohol-based mouth rinse (Biothene mouthwash, Laclede, Inc, CA) and a tooth paste with fluoride was recommended for daily use to promote remineralization and reduce risk of future caries. Good oral hygiene and diet were important components of the registered treatment plan. The patient was advised to adhere to low-salt, low-fat, low-calorie diet. He was also advised to increase consumption of potassium and protein-rich meals. The patient was called for control after a week following insertion of the restorations, and was recalled every 3 months afterward. The patient has been called for follow-up by both the dermatologist and the prosthodontist and has been fully satisfied and stable through the 1 year follow-up period free of new oral lesions (Fig. 4).

DISCUSSION

The typical clinical feature of PV consists of flaccid, thin-walled vesicles and bullae that arise usually on otherwise normal skin or mucosa but occasionally on erythematous skin. The lesions in the oral cavity are superficial and rupture easily, leaving painful, coalesced ulcers with ragged borders that gradually extend peripherally around teeth. It is difficult for patients to perform adequate oral home care because of pain and discomfort.[17] Also, there is an increased susceptibility to caries and periodontal disease.[18]

Medical management of PV involves topical, intralesional or systemic use of steroids and other immunosuppressive agents.[16] Sytemic corticosteroids are the first line of therapy for treatment of PV. When prednisone was introduced for treatment of PV, doses often were excessive (more than 200 mg/day) and were associated with considerable morbidity. Lower doses subsequently proved to be very effective and were associated with considerably less morbidity.[10] Chronicity and relapse are common and well-known features of these

Fig. 4: Intraoral views after 1 year follow-up

conditions.[16] With early diagnosis and aggressive treatment, between 50 and 80% patients with PV achieve complete remission.[10]

Chronic ulcerations (lasting more than 2 weeks) on any oral mucosal surface cannot be attributed to some other factor should prompt the clinician to include PV in the differential diagnosis. Often tissue fragility becomes overt in areas of trauma from tooth brushing or from frictional forces caused by removable prosthesis.[1] Hence, oral mucosa pain associated with dental treatment can be reduced by minimizing trauma.[9] Long-term success will depend upon the prevention of future dental disease or complications. The clinician should correct any potential sources of oral irritation such as rough restorations, ill-fitting dentures, traumatic oral hygiene procedures and dysfunctional oral habits. Wearing tissue-borne prosthesis can be difficult, if not impossible, with PV lesions.

Fixed prosthesis are preferred wherever possible and great care should be taken to create optimal gingival contours to prevent inflammation and facilitate hygiene,[9] as was performed in the presented case where fixed metal-reinforced porcelain restorations were preferred over a removable one in the mandibular arch.

To the authors' knowledge, there is no literature elicited related to PV patients in need of fixed prosthodontic treatment. The prepared margins on the teeth, thus the margins of the fixed restorations, could preferably be placed supragingivally or with level with the gingival margins, so as not to traumatize the tissue and to gain access for daily oral hygiene procedures to prevent caries and periodontal breakdown. The design of the pontics is also important. Modified ridge-lap pontics for anterior dentition, hygienic or modified hygienic pontics where esthetics is not important and conical pontics where there is a knife-edge residual alveolar ridge may be used. To enable optimal plaque control, the gingival surface must have no depression or hollow. Rather, it should be as convex as possible from mesial to distal aspects. The critical point is then, to provide pressure-free contact between the pontic and the underlying tissues, this will allow for passage of floss over the entire tissue surface. Highly glazed porcelain is generally considered the most biocompatible material therefore, for easier plaque removal and biocompatibility, the tissue surface of the pontic should be prepared in glazed porcelain.

It is also imperative to care extra attention when making impressions due to active vesiculobullous lesions. This is often overcome with patience, gentle oral manuplation and careful technique.[19] However, it may be necessary to handle these treatment phases under the suppression of drug therapy, with consulting the dermatologist.

Slow deposition of local anesthetics as well as the use of pediatric films for intraoral radiography are preferable, as they minimize lateral tissue distortion and subsequent precipitation of oral blisters. Small soft-bristled toothbrushes are especially useful for these patients. Bland, nonalcohol-based antimicrobial rinses and flavor-free oral hygiene products are preferable to protect teeth from caries and to prevent secondary infection or irritation of tissue erosions and ulcerations.[20,21] In addition, a high-concentration fluoride dentifrice may be advised to these patients.[9] Therefore, a nonalcohol-based mouth rinse and a tooth paste with fluoride was recommended for the patient presented. Optimal oral hygiene is important because the gingival involvement may represent an exaggerated response to bacterial plaque.[17] Maintenance of a healthy dentition is also essential for ensuring adequate nutrition through efficient

mastication. Diet counseling should emphasize the control of dietary cariogenicity while maintaining high-calorie nutrition required for tissue repair in these patients.[20]

Patients with PV should be receiving regular and frequent dental examinations (every 2 to 3 months) until extended intervals can be justified.[9] Therefore, the presented patient has been called for follow-up recalls every 3 months.

The most important aspect of PV is its early recognition, diagnosis and treatment. The dental clinician and healthcare team play a critical role in the quality of life for these patients. It is essential to monitor the patient in collaboration with a dermatologist. Though the dental problems in PV can be challenging, thoughtful and comprehensive dental treatment planning and execution will result in favorable outcomes. The patient in the presented case has tolerated his fixed prosthesis well and has been using them comfortably since the beginning of rehabilitation procedures.

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