

CLINICAL CASE

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GIUSEPPE ALESSANDRO SCARDINA¹, FRANCESCO CARINI², PIETRO MESSINA¹,
VINCENZO VALENZA², ANGELO LEONE²

Pemphigus Vulgaris in a Mediterranean Young Adult Manifesting as Desquamative Gingivitis

Pęcherzyca zwykła w postaci łuszczonego zapalenia dziąseł u chłopca z rejonu śródziemnomorskiego

¹ Department of Oral Science, University of Palermo, Italy

² Department of Experimental Medicine, Section of Human Histology and Anatomy, University of Palermo, Italy

Abstract

The authors present a rare case of pemphigus vulgaris in a 17-year-old Sicilian male patient manifesting as desquamative gingivitis. Clinical examination revealed erythematous gingival with ulcerated areas and evidence of bullae ruptures. White plaquelike lesions were also observed. Nikolsky sign was positive. There were no other mucosal or skin lesions. Gingival biopsy showed histopathological and immunological patterns typical of pemphigus vulgaris. The patient was treated with topical immunosuppressive therapy and the follow-up showed maintenance of oral health. This report underlines the essential role of the periodontist in the early diagnosis of a serious disease (Dent. Med. Probl. 2005, 42, 2, 367–369).

Key words: desquamative gingivitis, oral pemphigus vulgaris.

Streszczenie

Autorzy opisują rzadki przypadek pęcherzycy zwykłej u 17-letniego Sycylijczyka w postaci złuszczonego zapalenia dziąseł. Badanie kliniczne ujawniło zaczerwienienie dziąsła z nadżerkami oraz pękniętymi pęcherzami. Stwierdzono także białe wykwity tarczopodobne. Objaw Nikolskiego był dodatni. Nie występowały inne zmiany na błonie śluzowej oraz zmiany skórne. Badanie histopatologiczne i immunologiczne dziąsła wykazywało typowe zmiany dla pęcherzycy zwykłej. Pacjent był leczony miejscowo lekami immunosupresyjnymi i w obserwacji po badaniu nie stwierdzono nawrotów zmian w jamie ustnej. Opis przypadku wskazuje na istotną rolę periodontologa we wczesnej diagnostyce poważnie przebiegającej dermatozy (Dent. Med. Probl. 2005, 42, 2, 367–369).

Słowa kluczowe: złuszczone zapalenie dziąseł, pęcherzyca zwykła w jamie ustnej.

A 17-year-old Sicilian man was referred by his reliable dentist to Oral Medicine section, Department of Odontostomatological Science of Palermo for a definite diagnosis. He observed spontaneous bleeding from the gingival and painful desquamation. The patient had no notable medical, cutaneous, genital, or ocular problems. Extra-oral examination revealed no abnormalities. Oral examination revealed only gingival lesions with erythema and erosions of the attached and marginal gingiva (Fig. 1). Nikolsky's sign was positive (Fig. 2); associate symptoms were severe pain on contact with food and bleeding. This clinical picture suggested a vesiculobullous disorder (Fig. 3). Erosive

lesions were biopsied from the periphery of the lesion to ensure some attached epithelium for ideal histology and immunofluorescent analysis.

The result showed intense acantholysis in the lower stratum spinosum, with the basal cells of the epithelium staying adherent to the basement membrane, suggesting a diagnosis of pemphigus vulgaris (PV) (Fig. 4). Direct immunofluorescence showed IgG, IgM and C3 deposits in the stratum spinosum, thus confirming the diagnosis of PV.

The patient was treated with topical cyclosporine applications, 5 ml of oral suspension (500 mg), shared two times a day; cyclosporine had to be kept in the oral cavity for 15 minutes and

no food nor drinks were allowed for 30 minutes before and after the oral wash. The topical therapy resulted in a complete resolution in 3 months. No side effects have been reported and the monitoring of full blood count, of renal and hepatic function and of blood pressure has not shown any variation from the baseline.

Discussion

Chronic desquamative gingivitis (DG) is a common condition, which is mostly first recognized by periodontist. DG is a clinical manifestation of several different disease processes: Nisengard et al. showed that 48.9% of the cases of DG were due to cicatricial pemphigoid, 23.6% to lichen planus, and only 2.3% to pemphigus. Similar appearance may be seen in reaction to medications, dental materials, candidosis, lupus erythematosus, Chron's disease, sarcoidosis, leukaemias, chewing gum and mouthwashes [1].



Fig. 1. PV with the aspect of gingivitis: gingival lesions with erythema

Ryc. 1. Zmiany dziąsłowe w przebiegu pęcherzycy zwykłej: zmiany nadżerkowe z rumieniem



Fig. 2. Epithelial detachment with positivity of the Nikolsky sign

Ryc. 2. Złuszczenie się nabłonka dziąsłowego z dodatnim objawem Nikolskiego

Since DG is a common clinical presentation of a variety of diseases, definitive diagnosis and treatment are problematic. Therefore, a proper diagnosis of the underlying pathologic entity must be established in order to provide the appropriate therapy. DG is usually a manifestation of lichen planus or mucous membrane pemphigoid and may be the only or initial clinical presentation of these disorders. A variety of less common mucocutaneous diseases can also cause DG [2].

Pemphigus is the most serious cause, as it is potentially lethal. Pemphigus is a chronic bullous disease that involves the skin and mucous membranes. Although there are several forms of pemphigus, the most important one affecting mucous membranes is PV.

PV is characteristically seen in older patients and shows a predilection for the Jewish race and Mediterranean origin [3]. PV is an autoimmune disorder in which there is damage to desmosomas by antibodies directed against cadherin-type epithelial cell adhesion molecules, particularly Dsg3 [4].

Most PV cases develop intraoral lesions at the early stage, showing skin lesions later on. Oral lesions are common, the bullae quickly become erosive and are covered with necrotic material.



Fig. 3. Blisters show ruptures to leave flaps of peeling tissue with red erosions

Ryc. 3. Rozerwany pęcherz prowadzi do powstania czerwonej nadżerki

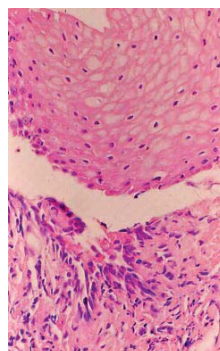


Fig. 4. Histological aspect: the epithelial blister

Ryc. 4. Zmiany histologiczne typowe dla pęcherzycy zwykłej i pęcherz nabłonkowy

The gingival is frequently involved and in some cases manifests as DG. Erosions heal slowly but scarring is rare. Gingival lesions usually include severe desquamative or erosive gingivitis, in which blisters show ruptures to leave flaps of peeling tissue with red erosions or deep ulcerative craters mainly on the attached gingivae. Nikolsky's sign is not specific for PV [2]. The symptomatology is often aggravated by the presence of dental plaque, so that efficient treatment is not possible unless the concurrent periodontal disease is also addressed.

Biopsy examination and immunological examination are essential to the diagnosis. Immunofluorescence has increasingly been used as regards routinary histology to reach a DG more accurately diagnosis [4]. Direct immunofluorescence shows predominantly IgG, and sometimes C3c, IgA, IgM deposits in the intercellular space in either the lesional epithelium and clinically heal epithelium adjacent to the lesions. Assay of serum antibody titers by indirect immunofluorescence may also help to guided prognostication and therapy [4].

Current treatment is largely based on systemic immunosuppression with systemic corticosteroid, using azathioprine, dapsone, methotrexate, cyclophosphamide and cyclosporine as adjuvants or alternatives. Plasmapheresis, sometimes combined with cyclophosphamide and extracorporeal photophoresis, is surely beneficial [3].

In the reported case, such pathology did not undergo an early diagnosis from the dentist, who treated PV as a periodontal effect of a normal disease. The periodontal signs are frequent and are direct consequence of an impossible oral hygienic norms actuation, because of the painful pathology. The persistence of clinical signs and symptoms persuaded the dentist to refer the patient to our university centre. The dentist must suspect the gingival involvement as prior region of pathology which may have a severe systemic complication. The correct and early diagnostic recognizing is essential for pathologies like PV prognosis. As many of the PV cases are first referred to periodontists or dentists, the authors believe that the present clinical report may be informative for making an early diagnosis of similar cases.

References

- [1] SCULLY C.: Mucocutaneous disorders. *Periodontology* 2000, 1988, 18, 81–94.
- [2] MIGNOGNA M. D., LO MUZIO L., BUCCI E.: Clinical features of gingival pemphigus vulgaris. *J. Clin. Periodontol.* 2001, 28, 489–493.
- [3] PACOR M. L., BIASI D., CARLETTO A., MALEKNIA T., LOMBARDO G., LUNARI C.: Topical cyclosporine in the treatment of oral pemphigus. *Min. Stomatol.* 1988, 47, 183–186.
- [4] YIH W. Y., MAIER T., KRATOCHVIL F. J., ZIEPER M. B.: Analysis of desquamative gingivitis using direct immunofluorescence in conjunction with histology. *J. Periodontol.* 1998, 69, 678–685.

Address for correspondence:

Giuseppe Alessandro Scardina
Department of Oral Science
Via del Vespro 129
90127 Palermo
Italy
fax: 39 091 655 22 02
e-mail: scardina@odonto.unipa.it

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