Coexistence of mucous membrane pemphigoid and vitiligo

Sanath Aithal, Satyaki Ganguly, Sheela Kuruvila

ABSTRACT

Mucous membrane pemphigoid describes a rare heterogeneous group of chronic, inflammatory, mucous membrane-dominated, subepithelial blistering diseases that manifest a varying constellation of oral, ocular, skin, genital, nasopharyngeal, esophageal, and laryngeal lesions. Life-threatening airway obstruction and sight-threatening ocular scarring can occur in this condition, which is rarely reported in Indian literature. Vitiligo is another acquired autoimmune disorder characterized by loss of melanocytes. Vitiligo is associated with a number of disorders also considered to be autoimmune. Here we report a very rare coexistence of MMP and vitiligo, the first such report from India.

Key words: Autoimmune, cicatricial pemphigoid, mucous membrane pemphigoid, ocular pemphigoid, vitiligo

INTRODUCTION

Mucous membrane pemphigoid (MMP) is a putative autoimmune, chronic inflammatory, subepithelial blistering disease predominantly affecting mucous membranes.[1] Various basement membrane zone components have been identified as targets of autoantibodies in MMP. Mucous membranes that may be involved include the oral cavity, conjunctiva, nasopharynx, larynx, esophagus, genitourinary tract and anal canal. Vitiligo is another clinically well-characterized, chronic autoimmune skin condition. The concomitant occurrence of these two entities in a single patient is rare.

CASE REPORT

A 37-year-old male patient presented with complaints of redness and pain in his left eye since two years, painful erosions in the oral cavity since six months, and ulcers over the scrotum and right knee since two months. On ocular examination, there was congestion of the left sclera mainly on the lateral side along with a fibrotic band connecting the upper palpebral conjunctiva with the bulbar conjunctiva near the lateral canthus [Figure 1]. On examination of the oral cavity there were widespread erosions involving the inner side of lips, gingival, and labial mucosa [Figure 2]. Detailed examination revealed a single depigmented macule over the glans penis [Figure 3], which according to the patient appeared insidiously around 8 years ago and remained static. Based on the clinical features, a diagnosis of mucous membrane pemphigoid was considered. A Tzanck smear from the oral erosions, done to rule out pemphigus vulgaris, failed to show any acantholytic cells. Subsequently, the patient developed multiple tense vesicles and bullae over his left ear. In addition there was involvement of nasal mucosa, larynx, scrotal skin, perianal skin, and right knee [Figure 2]. A skin biopsy from the right ear showed subepidermal bullae and fibrosis beneath it [Figures 4 and 5]. The direct immunofluorescence of perilesional uninvolved skin showed strong linear deposits of IgG along...
the basement membrane zone. Detailed hematological, biochemical, and imaging studies did not reveal any evidence of malignancy. The patient was placed on a combination regimen of oral corticosteroid and daily cyclophosphamide. The long-standing depigmented macule over the glans penis did not show any evidence of atrophy, ruling out lichen sclerosus et atrophicus. Hence it was diagnosed as mucosal vitiligo and the patient was counseled regarding it.

**DISCUSSION**

MMP is described as a heterogeneous group of chronic, inflammatory, mucous membrane-dominated, subepithelial blistering diseases that manifest a varying constellation of oral, ocular, skin, genital, nasopharyngeal, esophageal, and laryngeal lesions. Life-threatening airway obstruction and sight-threatening ocular scarring can occur in this condition, also known as cicatricial pemphigoid, benign MMP and incorrectly as ocular pemphigoid. Various basement membrane zone components have been identified as targets of autoantibodies in MMP. These include bullous pemphigoid antigen 1 (BPAG1, 230 kDa), bullous pemphigoid antigen 2 (BPAG2, 180kDa), laminin 5, laminin 6, α6-integrin subunit, β4-integrin subunit, collagen VII, and other proteins of unknown identity and/or function. Considerable variability exists in the clinical presentation of MMP.

Scarring is common at non-oral sites of involvement, contributing to disease-related morbidity. A multidisciplinary approach is essential in the management of MMP. Early recognition of this disorder and treatment may reduce disease-related complications. The choice of agents for treatment of MMP is based upon the sites of involvement, clinical severity, and disease progression.

Cicatricial pemphigoid (MMP) has been rarely reported in Indian literature. Nayar et al. found there is an association with autoimmune diseases, both organ and non-organ-specific in a group of 34 patients with cicatricial pemphigoid suggesting a possible genetic basis for the association.

Autoimmune mechanisms with an underlying genetic predisposition are the most likely causes of vitiligo, although neurohumoral and autocytotoxic hypotheses are alternative theories or contributing mechanisms. Vitiligo is associated with a number of disorders also considered to be autoimmune. Previously association of vitiligo with bullous pemphigoid either alone or along with other autoimmune diseases have been reported.

Gaspar et al. reported a case of a 68-year-old woman with vitiligo, primary hypothyroidism and cicatricial pemphigoid with severe laryngeal involvement necessitating tracheostomy. A thorough search failed to reveal any other report of co-existence of these two conditions in the published literature.

MMP is a rare, difficult to treat subepidermal blistering disorder. We believe that this is the first case report of vitiligo coexistent with mucous membrane pemphigoid from India. The
co-existence of these two conditions is probably is related to their common autoimmune etiology.

REFERENCES

1. Chan LS, Ahmed AR, Anhalt GJ, Bernauer W, Cooper KD, Elder MJ, et al. The first international consensus on mucous membrane pemphigoid: Definition, diagnostic criteria, pathogenic factors, medical treatment, and prognostic indicators. Arch Dermatol 2002;138:370-9.

2. Neff AG, Turner M, Mutusim DF. Treatment strategies in mucous membrane pemphigoid. Ther Clin Risk Manag 2008;4:617-26.

3. Ramanan C, Ghorpade A, Das MN, Bose U, Banerjee AK. Cicatricial pemphigoid: A case report. Indian J Dermatol Venereol Leprol 2001;67:212-3.

4. Nayar M, Wojnarowska F, Venning V, Taylor CJ. Association of autoimmunity and cicatricial pemphigoid: Is there an immunogenetic basis? J Am Acad Dermatol 1991;25:1011-5.

5. Deguchi M, Tsunoda T, Tagami H. Resolution of bullous pemphigoid and improvement of vitiligo after successful treatment of squamous cell carcinoma of the skin. Clin Exp Dermatol 1999;24:14-5.

6. Tirado-Sánchez A, Montes-de-Oca G. Coexistence of bullous pemphigoid, vitiligo, and thyroid disease: A multiple autoimmune syndrome? Dermatol Online J 2005;11:20.

7. Patel RS, Harman KE, Nichols C, Burd RM, Pavord S. Acquired haemophilia heralded by bleeding into the oral mucosa in a patient with bullous pemphigoid, rheumatoid arthritis, and vitiligo. Postgrad Med J 2006;82:e3.

8. Marcet B, Sibaud V, Géniaux M, Taieb A. Bullous pemphigoid, primary biliary cirrhosis and vitiligo: A multiple autoimmune syndrome? Ann Med Interne (Paris) 2002;153:349-50.

9. Pasić A, Ljubojević S, Lipozencić J, Marinović B, Loncarić D. Coexistence of psoriasis vulgaris, bullous pemphigoid and vitiligo: A case report. J Eur Acad Dermatol Venereol 2002;16:426-7.

10. Gaspar ZS, Wojnarowska F. Cicatricial pemphigoid with severe laryngeal involvement necessitating tracheostomy (laryngeal cicatricial pemphigoid). Clin Exp Dermatol 1996;21:209-10.

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