



Mucous Membrane Pemphigoid: A Frustrating Disabling Disease in A 15-year-old Male

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Abstract

Mucous membrane pemphigoid (MMP) is a rare chronic autoimmune subepithelial blistering disease characterized by erosive lesions of mucous membranes and skin that typically results in scarring of at least some sites of involvement. A progressive disorder that may result in serious complications including blindness, loss of the airway, and esophageal stricture formation. Most patient experience reduced quality of life specifically with ocular symblepharon and laryngeal destruction. Case: We present a case of a 15-year-old male patient with 1 month-history of multiple tense bullae, difficulty to open the eyes, and hoarseness due to scar lesion in mucous membrane. The patient was reported to have Dermatology Life Quality Index (DLQI) of 15 which implied he had severe disability and decreased quality of life due to his illness. Patient also had symblepharone release operation done by ophthalmologist. A laryngeal reconstruction plan was also carried out by ENT doctor, but the patient couldn't continue the treatment because of economic problem. Scarring of MMP is a significant complication in many cases. Mucous membrane pemphigoid could cause a lot of disability to the patient and decreased quality of life. Symblepharone release operation was done and laryngeal reconstruction plan was also carried out. Surgical intervention may not curable; however, it may be necessary for restoring function and improving quality of life.

Introduction

Mucous membrane pemphigoid (MMP) is a rare chronic autoimmune subepithelial blistering disease characterized by erosive lesions of mucous membranes and skin that typically results in scarring of at least some sites of involvement (Rashid et al., 2021). A rare disorder with a predominance in the elderly and an estimated incidence of 1 to 2 cases per million annually; females are affected 1.5 to 2 times as often as males (Kutlubay et al., 2021). Other resources said the prevalence of MMP is 5–7.5 cases per 10,000 adults, and the disease most often manifests in adults between 50 and 80 years of age (Al Ramil et al., 2023).

Lesions commonly involve the oral and ocular mucosae; other sites that may be involved include the nasopharyngeal, laryngeal, esophageal, and anogenital mucosae. A progressive disorder that may result in serious complications (eg, blindness, loss of the airway, esophageal stricture formation). Over the years, a variety of alternate designations, such as cicatricial pemphigoid, oral pemphigoid, desquamative gingivitis, ocular pemphigoid, ocular cicatricial pemphigoid, essential shrinkage of the conjunctivae, ocular pemphigus, have been applied to

MMP. Most patient experience reduced quality of life specifically with severe mucosal scarring including ocular symblepharon and laryngeal destruction (Hofmann et al., 2022; Ormond et al., 2020; Miyamoto et al., 2022).

Case Report

A 15-year-old male patient developed multiple mucocutaneous blisters on perioral, around the left eye, ears, neck, back, stomach, arms, legs, and groin that felt painful and increased in number within one month. Symptoms were initially presented as a bump inside the upper eyelid followed by redness of the eyes one year before the hospital admission. Within two weeks, he developed tense blisters around the mouth and groin. The blisters left abrasions on the skin when it ruptured. The blisters and redness of the eyes healed within months and left scars, causing difficulty to open the eyelid. He also complaint of oral thrush and abrasions which caused difficulty to eating.

Six months prior to the presenting features, tense blisters appeared on the shoulder, back, stomach, groin and legs, accompanied with painful bumps and redness in the eyes. The patient was treated with artificial tears, oral clindamycin and topical fusidic acid 2%. No improvement was documented. The blisters healed within months and left scars. The symptoms reappeared after 5 months. There was history of weight loss in the past 7 months and nasal discharge and epistaxis in the past 5 months. No drug and food allergies were reported.

In physical examination, the patient was in moderate illness and very underweight. On ocular examination, we found symblepharone and epiblepharone. Dermatology examination showed tense bullae, excoriation, red blackish crust, scars and inflamed scars on the perioral, ears, neck, back, stomach, arms, legs and groin regions (Fig. 1). Mucosal involvements in mouth, conjunctiva, and larynx were documented.

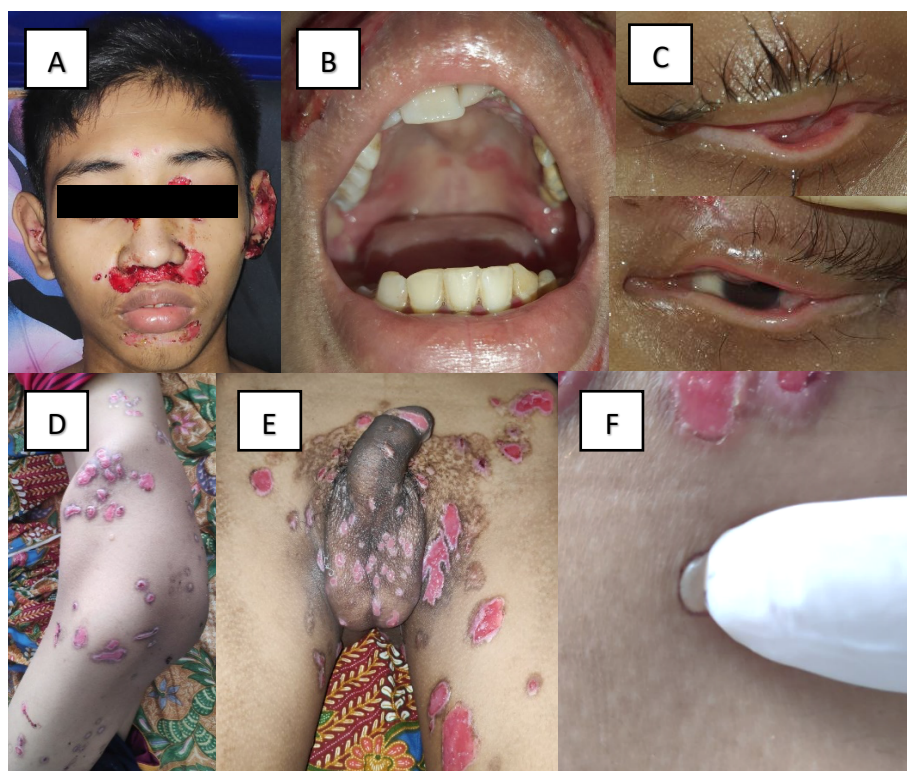


Figure 1. Clinical presentation. Skin manifestation in facial (A), extremity (D), and groin region (E). Oral (B) and ocular (C) mucosal involvement. Nikolsky's sign was negative (C).

The patient was reported to have Dermatology Life Quality Index (DLQI) of 15 (which implied he had severe disability and decreased quality of life due to his illness). Nikolsky's sign was negative. Laboratory investigations revealed anemia (Hb 8.6 g/dL), leukocytosis (leukocytes $12.43 \times 10^3/\mu\text{L}$), low levels of albumin and globulin (2.5 and 4.6 g/dL, respectively), and false-negative DIF. Skin biopsy examination was performed (Fig. 2). Indirect immunofluorescence (IIF) was not performed due to unavailability of the procedure in our center. Fiber optic laryngoscopy examination revealed there was laryngeal destruction with scar healing (Fig. 3). The patient also reported to had decreased of vision (right eye 3/60 and left eye 20/25).

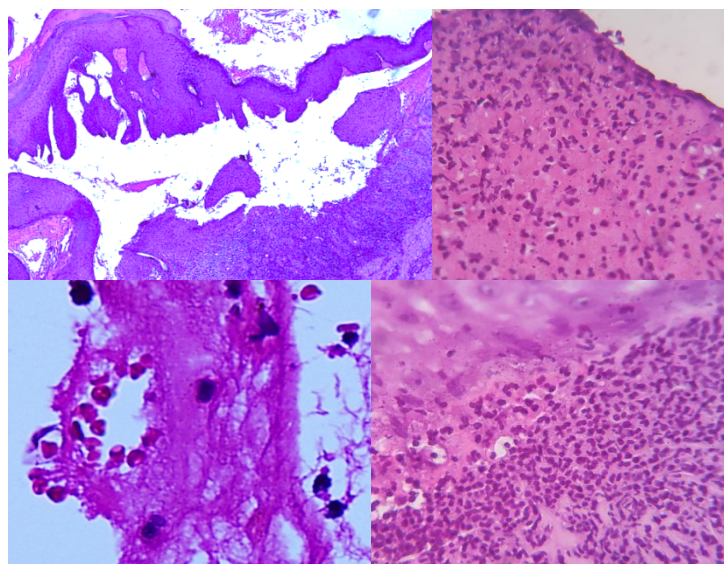


Figure 2. Skin biopsy.

Subepidermal bullae under the epidermis which separate the epidermis and dermis. In dermis and bullae cavity, there were infiltrate of neutrophils, lymphocytes, eosinophils and fibrins. The infiltrate in dermis suggested a neutrophil predominance. Neutrophil microabscesses were also found at the tip of the dermal papillae.

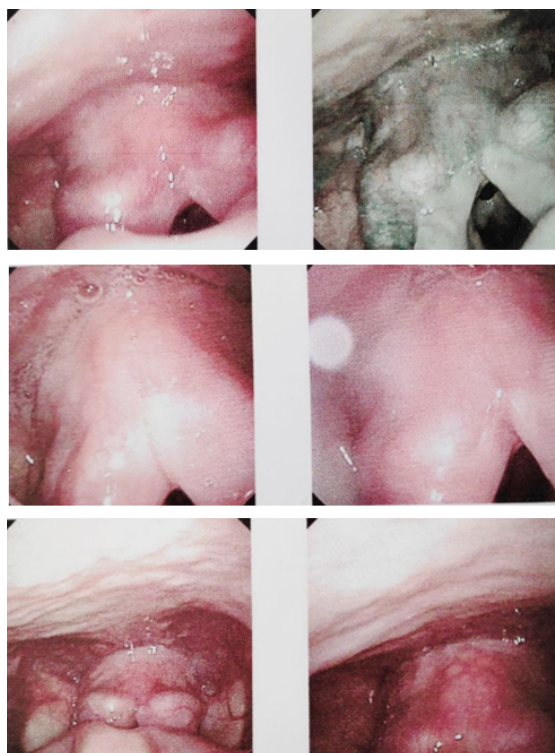


Figure 3. Fiber optic laryngoscopy.

Loss of omega shape of epiglottis, sinekia (+) on posterior arytenoid, symmetrical movement of plica vocalis with a bit of adhesion, rima glottis looked opened with minimal movement.

The patient was commenced with systemic therapy of oral prednisone 1 mg/kg/day with tapering off, ranitidine 150 mg twice daily, clindamycin 300 mg twice daily for 7 days, and mefenamic acid 500 mg 3 times daily. NaCl 0,9 % dressing was applied 3 times for 15 minutes on the crusts. Sodium fusidate ointment 2% was prescribed twice a day for the skin abrasion. After symptoms improvement, we continue to tapering off the dose of prednisone with additional treatment with azathioprine. Patient also had symblepharone release operation done by ophthalmologist. A laryngeal reconstruction plan was also carried out by ENT doctor, but the patient couldn't continue the treatment because of economic problem.

Result and Discussion

Discussion

We reported a 15th years old male with chief complaints of multiple tense blister that appeared on the perioral, around the left eye, both of ears, neck, back, stomach, both of arms, both of legs and crotch that felt painful and increased in number since 1 month ago. The patient also complaint of difficulties to open both of the eyes and hoarseness due to scar lesion in mucosal membrane. The patient diagnosed as suspect mucous membrane pemfigoid with differential diagnosis linear IgA dermatosis.

The diagnosis of suspect MMP is based on anamnesis, clinical features, tissue biopsy, and immunofluorescence examination. The most commonly affected sites in MMP include the oral cavity and eye, followed by the nasal, anogenital and pharyngeal mucosa and, less frequently, the oesophagus and larynx (Hofmann et al., 2022). Intraoral sites include the gingiva (80%), buccal mucosa (58%), palate (26%), alveolar ridge (16%), tongue (15%) and lower lip (7%) (Kumari et al., 2022). in MMP, blisters have a tendency to recur at the site of a previous lesion, scarring may be present in the dermis beneath a “new” blister.¹² In this patient, the mucosal involvement was in oral cavity, eyes, nasal, and larynx. In the anamnesis, some of the blisters reappeared on the scars tissue on the body. Most of the blister in the patient tends to leave scar after healed (Siriwardena et al., 2023).

We differentiated the diagnosis of this patient as linear IgA dermatosis based on clinical examination. There are tense bullae in patient's body, some of these lesion forming cluster of jewels pattern. Mucosal involvements could also be seen in patients with linear IgA dermatosis. Both MMP and LAD histopathological features are similar each other. Histology features of LAD are collection of neutrophils predominant at the basement membrane, often collecting in papillary tips with subepidermal blisters (Ajith Kumar, 2020; Mohanapriya, 2020). Histopathological features of MMP are subepidermal blister, viable or eroded roof over the blister, perivascular lymphocytes with variable eosinophils, neutrophils, or plasma cells if on mucous membrane. Depending on the age of the lesion, in lesions of less than 48 hours' duration, there are neutrophil microabscesses in the dermal papillae. With increasing age of the lesion, there are increasing numbers of eosinophils and later of lymphocytes and a reducing number of neutrophils (Mulder et al., 2021; Leal-Silva et al., 2022).

In this case, there is subepidermal bullae, in dermis and bullae cavity of the older lesion consist of infiltrate of neutrophil, lymphocyte, eosinophil and fibrin. The infiltrate in bullae cavity seems neutrophil predominant. In the newer lesion (<48 hours), the infiltrate in dermis consist of neutrophils and also neutrophil microabscesses at the tip of the dermal papillae. No eosinophil nor lymphocyte found in the newer lesion. These findings could be an aid to differentiate with other differential diagnosis.

Medical management for MMP are divided in group of “High-Risk” patients and “Low-Risk” patients. The participants define “high-risk” patients as those who have disease occurring in any of the following sites: ocular, genital, nasopharyngeal, esophageal, and laryngeal mucosae. For patients with severe disease or rapid progression of disease (particularly patients with ocular, esophageal, or laryngeal involvement) the initial treatment should be with prednisone (1-1.5 mg/kg per day) and cyclophosphamide (1-2 mg/kg per day) (Mei & Peters, 2021). In this case we give prednisone 1 mg/kg/day with tapering off as the treatment for the patient. Symblepharone release operation was done and laryngeal reconstruction plan was also carried out. Scarring is a significant complication in many cases. Surgical intervention may not curable; however, it may be necessary for restoring function and improving quality of life.

Conclusion

We reported a 15th years old male with chief complaints of multiple tense blister that appeared on the perioral, around the left eye, both of ears, neck, back, stomach, both of arms, both of legs and crotch that felt painful and increased in number since 1 month ago. The patient also complaint of difficulties to open both of the eyes and hoarseness due to scar lesion in mucosal membrane. This patient was diagnosed as MMP based on anamnesis, clinical features, and tissue biopsy. Multidisciplinary collaboration is often necessary for the diagnosis and proper treatment of MMP. Surgical intervention is not curable, however, it may be necessary for restoring function and improving quality of life.

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