International Journal of Research in Medical Sciences
Ramineni HB et al. Int J Res Med Sci. 2015 Jun;3(6):1543-1544
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: 10.18203/2320-6012.ijrms20150186

Case Report

Pemphigus vulgaris: a rare case report

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Received: 31 March 2015
Accepted: 07 May 2015

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ABSTRACT

Pemphigus Vulgaris (PV) is an organ-specific autoimmune disorder affecting skin and mucous membranes with a characteristic of intraepithelial blistering. The first common sign of this disease is involvement of oral mucosa followed by skin involvement. We here in report a rare case one such recently seen by us, where oral lesions and skin lesions in a 24 year old female patient presenting with a five months history of multiple fluid filled lesions all over the body, who is known case of psychosis finally diagnosed as having pemphigus vulgaris.

Keywords: Pemphigus vulgaris, Autoimmunity, Intraepithelial blisters, Oral lesions

INTRODUCTION

Pemphigus Vulgaris (PV) is an autoimmune disease affecting oral mucosal membrane and skin. This disease shows a characteristic feature of intraepithelial blistering of both the membranes. The first sign and part of involvement is oral mucosa in majority of patients and later it precedes the involvement of skin.[1] In some cases, oral lesions may also be followed by skin involvement. A recent study from the UK suggests an incidence of 0.68 cases per 100000 person years. Incidence varies in different parts of the world, being more common in the near and Middle East Europe than in Western Europe and North America.[2] All ages can be affected by Pemphigus, though it is most common in middle age, and both sexes are equally affected.[4]

CASE REPORT

A 24 year old female patient was presented with complaints of multiple fluid filled lesions all over the body since five months and followed by fever on and off with joint pains since ten days. Initial lesions are started from oral cavity for which medications are taken but not relieved. One month later these lesions spread all over the skin, for which patient was taking Prednisolone 100 mg for one week, later dose is reduced to 30 mg from last 20 days. She had a history of psychosis, and was on medication. She had altered sleep pattern and urinary incontinence. Her vital signs were temperature 103°F, pulse 140/minute, blood pressure 120/50, respiration 26/minute, and weight 35 kg, height 5'0". On dermatological examination, the patient was ill, moderately built and moderately nourished. Skin is characterized with multiple vesicles present all over the body (Figure 1), crushed lesions and large erosions are present over the thighs. Oral cavity is characterized with lesions. Routine laboratory examination showed Blood: ESR - 45 mm/hour, PCV - 24%, RBC - 2.9 mil/mm^3, Serum proteins - 4.3 g/dl, Albumin - 1.1 g/dl, Globulin - 3.2 g/dl, A/G ratio - 0.3.2. WBC, S. creatinine, Serum bilirubin, SGOT, SGPT, Alkaline phosphatase levels were within normal limits. Initially Patient was given

with inj. paracetmol 10 mg/ml, inj. cefoperazone sodium 5 gm TID, candid mouth paint, Tab. Fluconazole 150 mg OD, inj. albumin 100 ml infusion at rate of 10 ml/hour for 2days later prescribed with haloperidol ampoule IM OD, inj. dexamethasone 4gm 7 cc IV, 2 weeks of inj. optineuron (Vitamin B complex) IV, OD, later tab. prednisolone 30 mg and tab. Shelcal (Calcium carbonate + vitamin D_3) 500 mg, OD. tab. esomiprazole 40mg OD, tab. levocitrazine 5mg OD, tab. quitiapine 50 mg, T-Bact cream over the lesions for 2 weeks. On follow-up patient was symptomatic and responded well to the treatment.

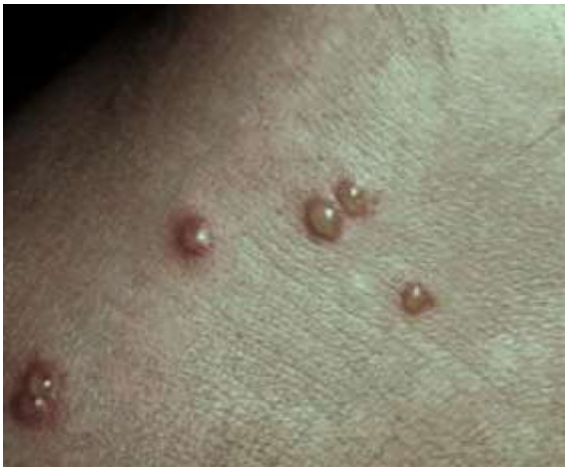

Figure 1: Multiple vesicles filled with clear fluid.

DISCUSSION

Pemphigus Vulgaris is a blistering disease of oral and skin mucosa membrane which is rare in Asia. The diagnosis of this disease is typical due to the appearance of lesions in the oral cavity followed by on skin which is still a mystery with a differential diagnosis along with Crohn's disease and vitamin B12 deficiency.[1,2,5] PV is an autoimmune disease affecting people in middle ages and distributed evenly in both sexes.[6] The initial lesions are often menacing and localized. Oral cavity is affected by persistent, painful ulcers with burning sensation that affects the appetite of the patient. The skin is affected after few weeks or months, with the appearance of blisters filled with clear fluid. These blisters are easily broken forming epidermal rings. Histological examination shows cleavage in the intraepidermal layer, with a basal cellular layers the base of the blister.[3] The basic therapy options for pemphigus consists of either local or systemic corticosteroid therapy.[4] In cases where PV is not extensive and lesions are limited local corticosteroid therapy can be initiated but oral corticosteroids shows adverse effects. Adjuvant therapies that are preferred include colchicine, thalidomide and retinoids shown to be beneficial in mild to moderate cases.[4,6] Recent trails suggest use of low-dose Methotrexate with more efficacy and no side effects.[6] Mycophenolate mofetil is a new drug with few side effects; it has spectacular effects when used in conjunction with corticosteroids. Some traditional drugs are combined along with systemic corticosteroid therapy, such as calcium and vitamin supplements, and gastric protecting agents.[7] Strict local hygiene should be maintained with a diluted antiseptic mouthwash to improve the patient's comfort.

CONCLUSIONS

Pemphigus vulgaris is a rare chronic autoimmune disorder affecting cutaneous-mucosa that is often misdiagnosed and late, even then oral lesions appear. If not treated promptly, the disease has a high morbidity rate, with a fatal rate of 5% to 10% in most of cases. The diagnosis is confirmed through physical and pathological examination. The therapeutic regimen, along with corticosteroid therapy and adjuvant treatments, helps patient to relive from symptoms.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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DOI: 10.18203/2320-6012.ijrms20150186
Cite this article as: Ramineni HB, Yerraguntla M, Pulimila S, Kukkapalli NB, Suryadevara V. Pemphigus vulgaris: a rare case report. Int J Res Med Sci 2015;3:1543-4.