Original Article

A Rare Association Of Iga Pemphigus With Supraglottic Squamous Cell Carcinoma – A Case Report

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Abstract

IgA Pemphigus is a very rare form of pemphigus characterized by pruritic vesiculopustular eruptions with autoantibodies directed against desmosomes of keratinocytes. The disease is commonly associated with monoclonal IgA gammopathy, B-cell lymphoma, Inflammatory bowel disease, HIV infection and thiol drugs. Here we describe a case of IgA pemphigus associated with supraglottic squamous cell carcinoma. Tzanck smear showed acantholytic cells. Skin biopsy showed intraepidermal blister with mixed neutrophilic and eosinophilic infiltrates. On performing enzyme linked immunosorbent assay (ELISA), IgA autoantibodies reacted to desmocollin 1 antigen. On video laryngoscopy, an ulceroproliferative growth of size 2.3 x 1.6 x 4 cm cm is seen obliterating the left pyriform fossa in the supraglottis. All above results were correlated and a diagnosis of IgA Pemphigus associated with Supraglottic Squamous Carcinoma was made. To the best of our knowledge, no similar association has been reported so far.

KEYWORDS: Carcinoma, Immunobullous disorders, Paraneoplastic disorders, Vesiculobullous disorders.

INTRODUCTION

IgA (immunoglobulin A) pemphigus is a very rare form of autoimmune, intra-epidermal blistering disorder characterized by the presence of vesiculopustular skin lesions which are intensely pruritic and show annular configuration with crusting. It comprises two types namely subcorneal pustular dermatosis type (SPD) which shows autoantibodies directed against desmocollin 1 and intraepidermal neutrophilic type (IEN) for which the target antigen is uncertain. Several associations with other diseases such as monoclonal IgA gammopathy, B-cell lymphoma, Inflammatory bowel disease, HIV infection and thiol drugs have been made in the past. To the best of our knowledge, we report this as the first case of IgA Pemphigus associated with Supraglottic squamous carcinoma.

CASE REPORT :

A 55-year-old male presented with complaints of multiple intense itchy fluid filled lesions over the abdomen, flanks, back, arms, forearms, thighs and legs for the past 15 days. The fluid filled lesions remained intact for over a week and only a few lesions ruptured. The patient also had a history of difficulty in swallowing (Solids > liquids) for the past 1 month along with change in voice and foreign body sensation. The patient also noticed loss of weight and loss of appetite for the past 1 month. Patient had no history of fever, joint pain, malaise, drug ingestion or topical application prior to the appearance of lesions.

On local examination, multiple erythematous plaques of varying sizes, smallest measuring 1 x 1.5 cm and largest measuring 10 x 15 cm, some with central hyperpigmentation present over abdomen, flanks, back, arms, forearms, thighs and legs. Multiple clear fluid filled vesicles, bullae and few pustules of varying sizes present over both clear and erythematous base, distributed over abdomen (Figure 1), arms and forearms (Figure 2). The bullae showed an annular pattern around erythematous plaques - string of pearls appearance. Multiple crusted

desquamations were noted around the annular plaques. The Nikolsky sign was negative. Oral mucosa did not show any lesions. Other general and systemic examinations were normal.

Complete blood count showed eosinophilia, ESR was mildly elevated. Tzanck smear showed acantholytic cells (Figure 3). Biopsy was taken from a recent lesion on the right forearm and showed multiple intraepidermal blisters with mixed neutrophilic and eosinophilic infiltrates (Figure 4 and 5). Enzyme linked immunosorbent assay (ELISA) showed antibodies reacting to desmocollin 1 antigen (anti-Dsc 1 autoantibodies). On video laryngoscopy, an ulceroproliferative growth of size 2.3 x 1.6 x 4 cm is seen obliterating the left pyriform fossa in the supraglottis (Figure 6). Biopsy was done for the same and showed features suggestive of Supraglottic Squamous Cell Carcinoma. Based on the above feature a diagnosis of Intraepidermal neutrophilic type of IgA pemphigus was made.

Patient was started on Inj.dexamethasone 1 cc(4mg) once daily and 100 mg dapsone once daily. Lesions resolved rapidly within a week. Patient was then maintained on oral corticosteroids and oral dapsone and referred to surgical oncology for further management.

Figure 1: Multiple clear fluid filled vesicles, bullae and few pustules of varying sizes arranged in annular configuration over and erythematous base in abdomen.

Figure 2: vesiculopustules arranged in annular configuration with central hyperpigmentation over left forearm

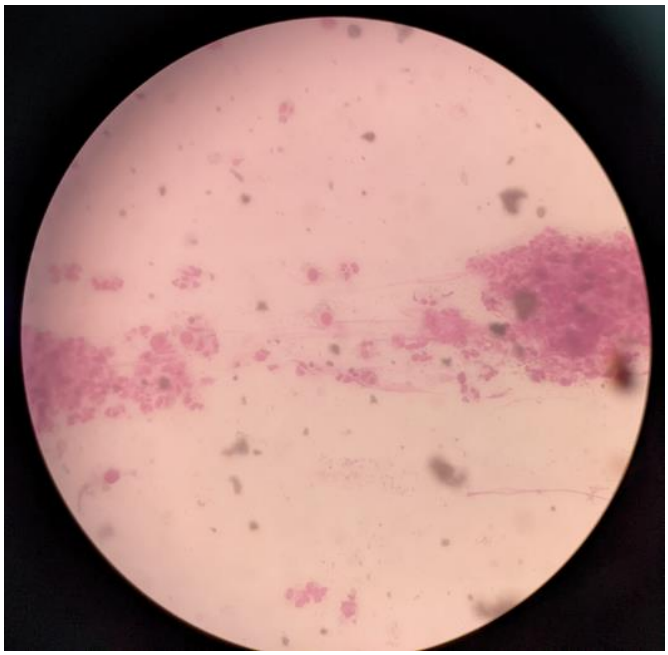

Figure 3: Tzank smear showing acantholytic cells

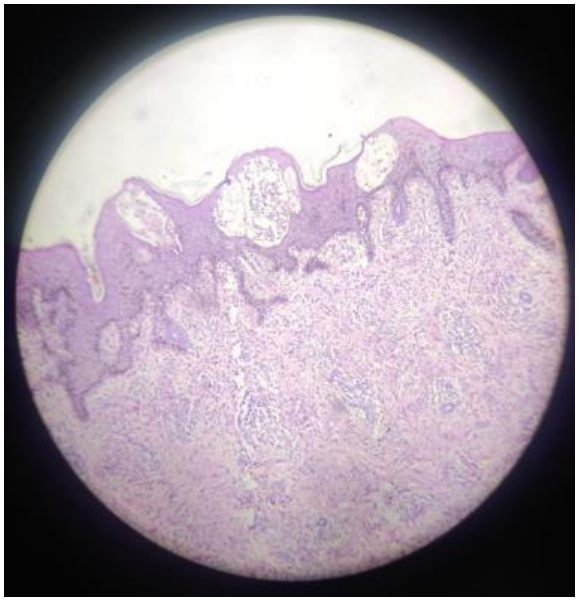

Figure 4: (H&E section, 10X magnification) skin biopsy showing intraepidermal blisters

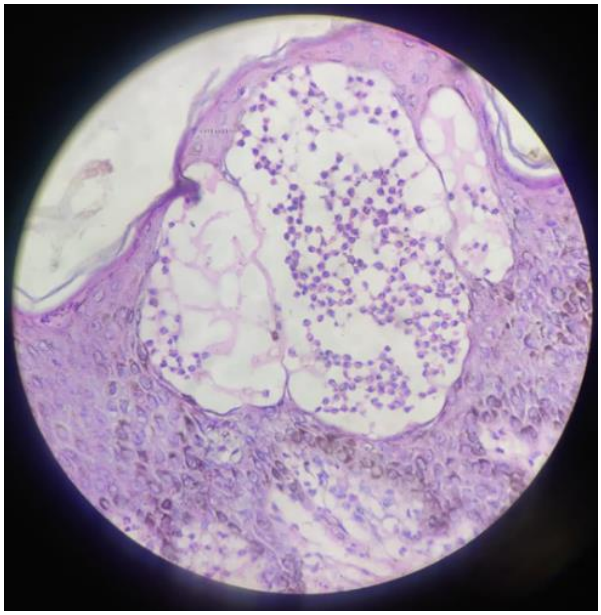

Figure 5: (H&E section, 40X magnification) Skin biopsy showing intraepidermal blisters with mixed neutrophilic and eosinophilic infiltrates

Figure 6: Video Laryngoscopy showing an ulceroproliferative growth of size 2.3 x 1.6 x 4cm cm seen obliterating the left pyriform fossa in the supraglottis

DISCUSSION:

IgA Pemphigus is a very rare autoimmune immunobullous diseases manifesting as vesiculopustular lesions with autoantibodies directed against epidermal cell surface antigens [1,2].Subcorneal pustular dermatosis (SPD)-type and intraepidermal neutrophilic type (IEN) are the two major types of IgA Pemphigus. There are also other minor subtypes such as IgA pemphigus vegetans, IgA pemphigus vulgaris, IgA pemphigus foliaceus and undetermined-type IgA Pemphigus [1,2,3]. SPD-type IgA Pemphigus is clinically characterized by superficial pustules in the intertriginous regions and IEN-type IgA Pemphigus by atypical pustular skin lesions. Diagnosis is usually made by histopathology showing intraepidermal neutrophilic pustule formation and immunofluorescence showing bound and/or circulating IgA cell-surface anti keratinocyte autoantibodies [3,4]. In SPD-type IgA Pemphigus, histopathology shows subcorneal neutrophilic pustules. In IEN-type histopathology shows neutrophilic pustules in the middle of epidermis. Reactivity to autoantigens such as desmogleins (Dsg), desmocollins (Dsc) and cell-to-cell adhesion molecules (cadherins) can be demonstrated by newer biochemical and molecular biologic methods [5,6]. Oral dapsone and systemic corticosteroids are the main treatments of choice. Other treatment options are tetracycline, colchicine, immunosuppressive agents, retinoids, adalimumab, plasmapheresis, and PUVA therapy[1,3,7].

IgA pemphigus has been reported with myeloma and B-cell lymphoma in the past [7,8]. Also, several types of solid organ neoplasms including lung carcinoma, ovarian carcinoma, carcinoma of gallbladder, adenocarcinoma in situof the pancreas, endometrial carcinoma and thymoma are reported in association with IgA Pemphigus [5,6,9,10]. However, IgA Pemphigus is being reported for the first time in association with Supraglottic Squamous Cell Carcinoma as in our case.

CONCLUSION:

We hereby report a case of IgA Pemphigus with its rare association with Supraglottic Squamous Cell Carcinoma. The use of dapsone with or without systemic corticosteroids remains mainstay of treatment. We also emphasize the need for screening all IgA pemphigus patients for any hidden malignancy.

REFERENCES:

1. Kanwar AJ, De D. Pemphigus in India. Indian J DermatolVenereolLeprol 2011;77:439-449 Robinson ND, Hashimoto T, Amagai M, Chan LS. The new pemphigus variants. J Am AcadDermatol 1999; 40: 649–671.
2. Grando SA. Pemphigus in the 21st century. New life to an old story. Autoimmunity 2006;39:521-30.

3. Niimi Y, Kawana S, Kusunoki T. IgA pemphigus: a case report and its characteristic clinical features compared with subcorneal pustular dermatosis. J Am AcadDermatol 2000; 43: 546–549.
4. Miyagawa S, Hashimoto T, Ohno H, Nakagawa A, Watanabe K, Nishikawa T, et al. Atypical pemphigus associated with monoclonal IgA gammopathy. J Am AcadDermatol 1995;32:352-7.
5. Reichel A, Benoit S, Giner T, Schmidt E, Goebeler M, Stoevesandt J. Anti-desmoglein 1 IgG/IgA pemphigus associated with thymoma. J DtschDermatolGes 2017;15:1147-8.
6. Porro AM, Caetano Lde V, MaeharaLde S, Enokihara MM. Non-classical forms of pemphigus: Pemphigus herpetiformis, IgA pemphigus, paraneoplastic pemphigus and IgG/IgA pemphigus. An Bras Dermatol 2014;89:96-106.
7. Sehgal VN. Pemphigus- A reappraisal of clinical and histological features in Indian patients. Dermatologica 1969;139:381-9.
8. Watkins C, West C, Kosari P, Ali S, Sangüeza O, Huang W. IgG/IgA pemphigus: Report of a rare variant of atypical pemphigus and a review of the literature. J DermatologClin Res 2014;2:1011.
9. Hashimoto T, Komai A, Futei Y, Nishikawa T, Amagai M. Detection of IgA autoantibodies to desmogleins by an enzyme-linked immunosorbent assay. The presence of new minor subtypes of IgA pemphigus. Arch Dermatol 2001; 137: 735–738.
10. Yasuda A, Kobayashi H, Hashimoto T, Itoh K, Yamane M, Nakamura J. Subcorneal pustular dermatosis type of IgA pemphigus: demonstration of autoantibodies to desmocollin-1 and clinical review. Br J Dermatol 2000; 143:144–148