Clinical Case Reports International

Case Report
Published: 23 Dec, 2020

Unrecognized Cicatricial Pemphigoid with Skin and Multiple Mucosal Complications - A Case Report

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Abstract

Cicatricial pemphigoid is a rare, chronic autoimmune bullous disease which resulted in scarring of mucosa predominantly conjunctival and oral mucosa. Uncommonly, the upper aerodigestive tract is involved and rarely skin is involved. We report an elderly Libyan woman with unrecognized cicatricial pemphigoid for 15 years; her disease was complicated by scared skin, mucosa of eyes and upper digestive tract.

Introduction

Cicatricial Pemphigoid (CP) is a rare chronic subepidermal blistering disease affecting mucous membranes and occasionally the skin. It may cause potentially life-threatening complications. The majority of patients present with oral erosions or desquamative gingivitis. Typical ocular lesions include chronic scarring conjunctivitis with progressive subconjunctival fibrosis, fornix foreshortening and synechia formation. The scarring can be a source of significant morbidity because it can result in odynophagia with eating and drinking difficulty and can lead to eventual blindness [1,2].

Case Presentation

An 84-year-old woman was admitted to our department because of localized blistering skin lesions around the umbilicus for many years associated with generalized pruritus. The patient was on liquid meals for long time as she had severe dysphagia. On examination; scarred plaque was seen in scalp (Figure 1), right shoulder and periumbilical area with few vesicles and erosions were seen around umbilicus (Figure 2). Multiple scratch marks were seen at discrete sites of skin (Figure 3). Desquamative gingivitis and multiple oral ulcerations were evident (Figure 4). Anogenital area was free. Ophthalmological examination revealed dry eyes, conjunctivitis with raw areas, fornix shortening, symblepharon, entropion and trichiasis (Figure 5), with corneal neovascularization.

Routine investigations were within normal apart from elevated blood sugar. Barium swallow study revealed free flow of barium throughout esophagus with two fixed narrow segments at cervical and upper thoracic esophagus (Figure 6). Periumbilical skin biopsy showed a subepidermal blister with many neutrophils and eosinophils with dermal scarring (Figure 7). The patient was responded to topical potent steroids with 0.5 mg/kg daily oral prednisolone; skin and mucosal erosions was healed but unfortunately, the patient was diagnosed in the advanced stage when scarring had already occurred.

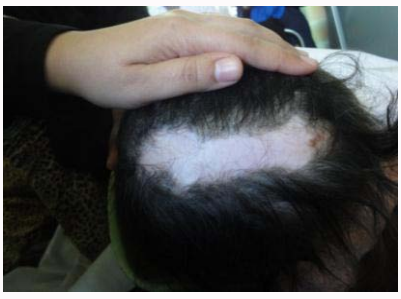

Figure 1: Scarring of scalp.

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Received Date: *18 Nov 2020*
Accepted Date: *17 Dec 2020*
Published Date: *23 Dec 2020*

Citation:
Elfaituri S, Twir I, Bheh A. Unrecognized Cicatricial Pemphigoid with Skin and Multiple Mucosal Complications - A Case Report. Clin Case Rep Int. 2020; 4: 1205.

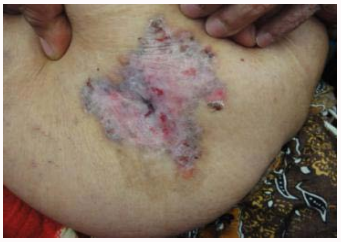

Figure 2: Periumbilical scarring with vesicles and erosions.

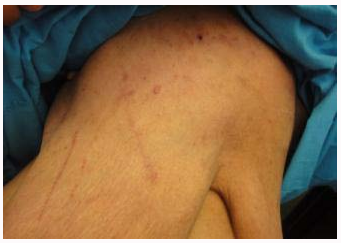

Figure 3: Multiple scratch marks due to pruritus.

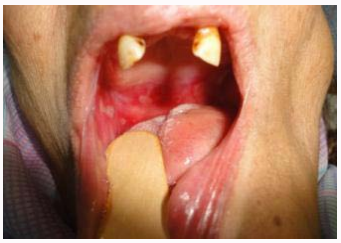

Figure 4: Oral ulcerations.

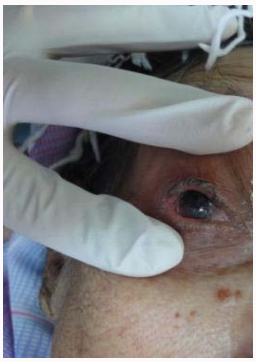

Figure 5: Entropion and trichiasis.

Discussion

CP, an autoimmune vesiculobullous disease commonly seen in the elderly is characterized by involvement of the mucous membranes with or without skin involvement. Oral involvement is the most consistent feature of CP, it presents with painful erosions and desquamative gingivitis resulting in scar formation of oral mucosa. Typical ocular lesions include chronic scarring conjunctivitis with progressive subconjunctival fibrosis, fornix foreshortening and synechia formation between the bulbar and palpebral conjunctiva [3]. Later, patients develop entropion with subsequent trichiasis, ocular surface keratinization, and eventually blindness [4]. Mucosal surfaces of pharynx, nasal mucosa, and oesophagus can also be affected [3]. Skin lesions develop in approximately one third of patients with

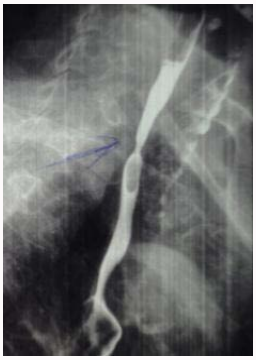

Figure 6: Narrowing of esophagus.

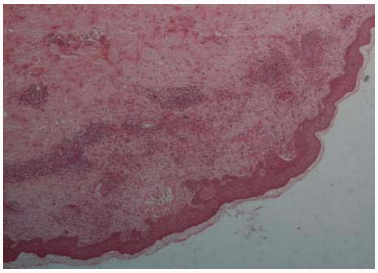

Figure 7a: Subepidermal blister with dermal scarring.

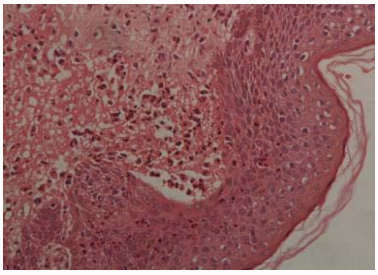

Figure 7b: Subepidermal blister with many neutrophils and eosinophils.

CP. Pruritus at the sites of blisters or generalized pruritus may be present. Blisters heal with scarring or milia. Scalp involvement lead to scarring alopecia. Other complications of CP include airway stenosis, esophageal stricture. Diagnosis is based on clinical and pathological findings and the detection of linear deposits of IgG and/or IgA and/or C3 at the dermal-epidermal junction by direct immunofluorescence microscopy of a perilesional biopsy [5]. Our patient presented with clinicopathological features of CP; however, there were several peculiar findings in this case, including delayed diagnosis for more than 15 years, association with generalized pruritus, esophageal scarring and scaring alopecia in addition to presentation with Periumbilical vesiculobullous lesions which are also typical for patients with classical bullous pemphigoid. Unlike the later condition, association with atrophic scarring was seen in our patient. The classical oral lesions as desquamative gingivitis and multiple oral ulcerations as well as eye findings as fornix shortening, symblepharon, entropion and trichiasis were typically seen in our case. Periumbilical skin biopsy showed a subepidermal blister with many neutrophils and eosinophils with dermal scarring which was consisting with CP. Immunoflorecence was not done as it was not available. Patients with mild localized CP may benefit from topical steroids whereas patients with more extensive disease and progressive scarring require systemic

steroids. Dapsone, immunosuppressive drugs, such azathioprine have also been used [5]. Our patient responded well to oral steroids. Unfortunately, the patient was diagnosed in the advanced stage when scarring had already occurred.

Conclusion

Cicatricial pemphigoid is a chronic progressive systemic autoimmune disease that has serious consequences. Recognition and early diagnosis is important to avoid scarring and improve the future prognosis of the disease.

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