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A CASE REPORT OF VESICULOBULLOUS ERUPTION IN A YOUNG PATIENT- DIAGNOSTIC DILEMMA AND THERAPEUTIC CHALLENGE

Dermatology

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ABSTRACT

Autoimmune bullous disorders are primarily classified as intraepidermal and subepidermal disorders. Subepidermal group of disorders comprises of many conditions like bullous pemphigoid, bullous systemic lupus erythematosus, linear IgA bullous dermatosis and epidermolysis bullosa acquisita. Bullous pemphigoid is the most common subepidermal disorder usually occurring in elderly individuals without any gender predilection. Its occurrence in adolescents appears to be even rarer. A 19-year-old female presented with multiple vesicles and bullae arranged in an annular pattern on face, trunk, bilateral upper limb and lower limb with mucosal involvement. Characteristic histopathology followed by direct immunofluorescence clinched the diagnosis of bullous pemphigoid. Although bullous pemphigoid is considered as a disease of elderly, it should be considered in the differential diagnosis of young patients presenting with itchy, tense blisters.

KEYWORDS

subepidermal bullous disorder, bullous pemphigoid, Linear IgA bullous dermatosis.

INTRODUCTION: Autoimmune bullous disorders are primarily classified as intraepidermal and subepidermal disorders. Bullous pemphigoid is an acquired autoimmune blistering condition characterized by formation of subepidermal blisters and accumulation of complement and antibodies along basement membrane zone. It primarily affects elderly, typically during Eight decade of life with no gender predilection. In Asia, incidence is estimated between 2.6-7.5 cases/million.(1) An Israeli study noted that younger bullous pemphigoid patients with mucosal involvement tend to have more extensive disease and require higher corticosteroid doses.(2) An Indian study reported average incidence age of 62.5 years.(3) It is rare among adolescents.(4) Bullous pemphigoid in adolescence appears even rarer than in early childhood.(5)

CASE PRESENTATION : A 19 year old girl presented with multiple itchy, reddish-raised lesions since one month and fluid filled lesions since one week. One month back she experienced severe itching starting from web spaces of fingers and spreading to whole body. Fifteen days later she developed itchy reddish-raised lesions for which she visited private practitioner and received anti-scabies treatment with no relief. A week later, she developed oral intolerance to spicy food followed by itchy, fluid-filled lesions starting from face. She was prescribed antiviral medications without relief and subsequently presented to us. The patient gave history of exacerbation of symptoms on sun exposure accompanied by burning micturition but denied history of preceding fever, trauma, radiation exposure or drug intake. There was no significant family history present.

General and systemic examination were unremarkable. Dermatological examination showed multiple tense vesicles and bullae with clear fluid arranged in annular pattern peri-orally, peri-umbilically, involving trunk, upper limbs and lower limbs with multiple discrete tense bullae as well. Crusted erosions on bilateral cheeks and nose were present. (Figure 1 is about here) Few macules on both palms were seen. Erosions over bilateral buccal mucosae and vulva along with tense bullae in nasal cavity were noted. Positive Bulla spread sign with negative Nikolsky sign were elicited while Tzanck smear was non-contributory.

Differentials considered were Bullous lupus erythematosus (on the basis of young age, history of photosensitivity, lesions on malar area), Linear IgA bullous dermatosis considering the “string of pearls” appearance and mucosal involvement, and bullous pemphigoid.

Laboratory investigations showed leukocytosis (total leucocyte count 32,000 cells/cumm), elevated absolute eosinophil count (10,240 cells/cumm) and elevated absolute neutrophil count (16,000 cells/cum). Antinuclear antibody test (ANA) by immunofluorescence technique was negative. Rest of the laboratory and serological investigations were normal.

Histopathology showed subepidermal split with infiltrate of neutrophils, eosinophils. Direct immunofluorescence from perilesional skin showed linear IgG and C3 along epidermal basement membrane with negative IgA, IgM, confirming diagnosis of bullous pemphigoid. (Figure 2 is about here)

She was started on intravenous dexamethasone 8mg/day, systemic antibiotics, Doxycycline 100 mg twice/day, Tab Dapsone 100 mg once/day and local care. After 15 days, due to fall in haemoglobin, dapsone was stopped. Despite treatment, 70-100 new crops of blisters continued to appear daily. (Figure 3 is about here) Hence, injection Rituximab (1gm) was started, but the infusion had to be abandoned due to tachycardia. Hence Tablet Azathioprine 50 mg twice/day was added to prednisolone in tapering doses with monitoring of complete blood count, liver function tests. The complete resolution of lesions was seen in one month. Patient has been following up with us since over a year with no recurrence of the lesions. (Figure 4 is about here)

DISCUSSION :

Bullous pemphigoid (BP) is a subepidermal blistering disorder with autoantibodies against Bullous Pemphigoid Antigen 230 (BP230) and Bullous Pemphigoid Antigen 180 (BP180) in hemidesmosomes of skin and mucous membranes. It is the most common autoimmune blistering disease globally, with annual incidence between 2.4- 21.7 cases/million.(6) BP is rare in younger individuals, with incidence below 0.5/million(7) It is more frequently drug-induced (induced by agents such as Dipeptidyl peptidase-4(DPP4) inhibitors, Penicillin, Penicillamine, Furosemide, Angiotensin converting enzyme (ACE) inhibitors, Antidiabetics, Nonsteroidal anti-inflammatory drugs

(NSAIDS) etc) in younger age groups but our patient denied consumption of any drug.

- Pathogenesis involves autoantibodies against BP230 and BP180 (collagen type XVII). BP180's major pathogenic epitope is the Non collagenous extracellular domain (NC16A) while BP230 is a cytoplasmic plakin protein. These antibodies then activate the complement pathway at dermo-epidermal junction leading to inflammatory responses (particularly eosinophils) culminating in blister formation. The disease presents with initial prodromal phase (itching, urticarial plaques) as seen in our patient. The bullous stage presents with tense vesicles and bullae on normal or erythematous plaques with negative Nikolsky sign.

- Clinical variants are dyshidrosiform, erythrodermic, papular, vesicular, toxic epidermolysis-like, lymphomatoid papulosis-like, ecthyma gangrenosum -like,localised, non-bullous. Mucosal involvement (seen in 20% patients) was an unusual manifestation in our patient with nasal, vulval mucosal involvement..(8) Multiple triggers like drugs, infections, vaccination, radiation, electric burns, radiotherapy are reported.(9)

- Clinical differential diagnosis of bullous pemphigoid include linear IgA disease, bullous lupus erythematosus, mucous membrane pemphigoid, epidermolysis bullosa acquisita (EBA). As patient had typical lesions on malar area and considering her young age, diagnosis of bullous lupus erythematosus was thought of. But it was ruled out due to negative ANA test and absence of features of interface dermatitis. Mucous membrane pemphigoid was ruled out because of lack of scarring. EBA was ruled out on the basis of age, absence of scarring, milia and blisters not only limited to trauma prone sites. Hence differentials were narrowed down to Linear IgA bullous dermatosis and bullous pemphigoid.

- Characteristic Histopathology shows subepidermal blister with eosinophils, neutrophils infiltrate. Histopathology alone cannot differentiate BP from other sub-epidermal blistering disorders. So, direct immunofluorescence was performed. Direct immunofluorescence demonstrates linear IgG and complement along the BMZ but sometimes IgA, IgE, IgM can also be present. Indirect immunofluorescence shows antibodies binding epidermal side of salt-split normal human skin. Due to non- availability, it was not performed in our case.

- Systemic corticosteroids are considered as gold standard for bullous pemphigoid. European Dermatology Forum guidelines suggest starting prednisolone 0.5–0.75 mg/kg/day for extensive and 0.5 mg/kg/day for mild disease. Topical clobetasol propionate 0.05% cream -30–40 g/day for extensive (excluding the face) and 20 g/day for mild cases.(10) Steroid sparing agents like Dapsone, Azathioprine, Mycophenolate mofetil, Cyclophosphamide, Tetracyclines, and Nicotinamide with general care are recommended. Our patient responded satisfactorily to combination of systemic and topical steroids with Doxycycline and Azathioprine.

- Zhou et al. have reported a 17 year old girl with recalcitrant bullous pemphigoid who demonstrated gradual improvement following a combination therapy consisting of injection Rituximab, Intravenous immunoglobulin (IVIG) and injection Dupulimab.(11) Similarly, Giacaman et al described a 17 year old male with generalised bullous pemphigoid with oral mucosal involvement as seen in our case who was managed effectively with systemic prednisolone.(4) Additionally, a case from India documented an 18 year old male with bullous pemphigoid and oral involvement, successfully treated using a combination of prednisolone, Doxycyline and Nicotinamide. Our patient was treated with Prednisolone in tapering doses and Tablet Azathioprine with complete clinical resolution of lesions in one month.

- Although Bullous pemphigoid is considered as a disease of elderly, it should be considered in differential diagnosis of young patients presenting with itchy, tense blisters. This case highlights unusual presentation of bullous pemphigoid at an unusual age.

FIGURES:

Figure 1: Multiple tense vesicles and bullae arranged in an annular pattern peri-umbilically, on upper limbs. Crusted erosion on bilateral cheeks with tense bulla in nasal cavity

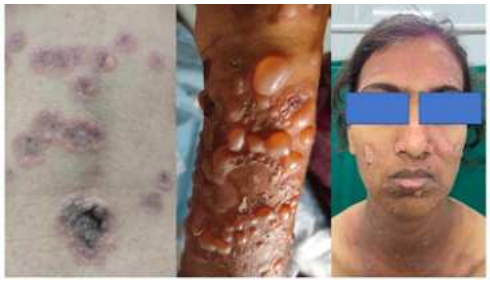


Figure 2: H & E 100x (Left)- Subepidermal split (Black arrow) Lumen of blister-inflammatory infiltrate of neutrophils, eosinophils, plasma cells.

Direct immunofluorescence (Right) - Linear positivity for IgG and C3 along epidermal basement membrane. Negative IgA and IgM

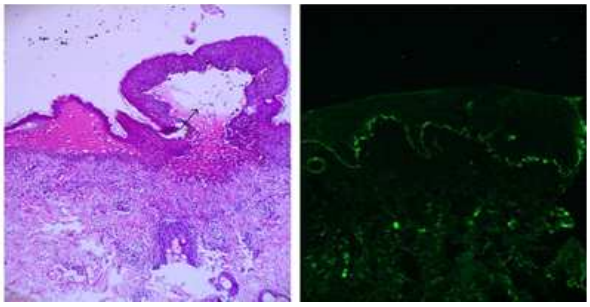


Figure 3: Multiple new vesicles and bullae, around 70-100 despite the treatment

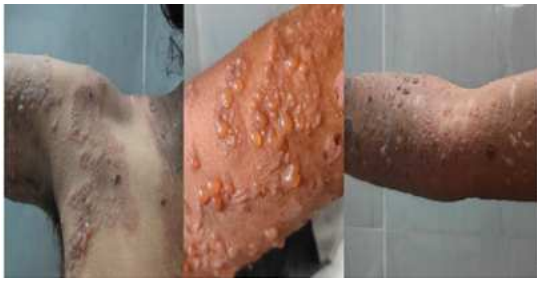
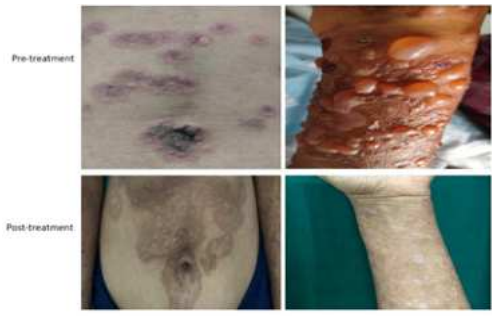


Figure 4: Pre-treatment and post-treatment clinical photos



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