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Polycyclic annular presentation of pemphigus vulgaris with an eosinophil predominance in two pregnant patients

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Abstract

Pemphigus during pregnancy has a more complicated course owing to the limitations in treatment options and alterations in the severity and presentation of the clinical features. We would like to present two pemphigus vulgaris (PV) cases associated with pregnancy with an unusual clinical appearance exhibiting polycyclic, annular, vesiculobullous plaques with marked eosinophil infiltration in histopathology. To the best of our knowledge pregnancy-associated pemphigus cases with this particular clinical presentation have not been reported in the literature. Changes in the immunologic and hormonal state during pregnancy may play a role in altering the classic clinical presentation and treatment response of PV.

Keywords: atypical, pemphigus vulgaris, pregnancy, polycyclic, annular

Introduction

Pemphigus vulgaris (PV) is an autoimmune bullous disease presenting with fragile blisters in the skin and mucous membranes and is potentially life threatening if left untreated [1]. Pemphigus during pregnancy has a more complicated course owing to the limitations in treatment options [2, 3]. A majority of the reported pemphigus cases showed exacerbation during the first and second trimesters of pregnancy and the post-partum period [4, 5]. We present two PV cases associated with pregnancy presenting with an unusual polycyclic annular distribution and marked eosinophil predominance upon histopathological examination.

Case Synopsis

Case 1: A 23-year-old woman presented with widespread vesiculobullae with severe pruritus and oral erosions during the 8th week of gestation. On physical examination severe erosions in the oral mucosa and blisters on the periphery of resolved lesions in a rosette-like fashion involving the trunk and extremities were observed (**Figure 1A-C**). Histopathological examination revealed mild acanthosis and spongiosis in the epidermis, intraepidermal eosinophils, mild edema in the papillary dermis, intensive perivascular infiltration with eosinophils, and a tendency toward subepidermal splitting (**Figure 1D, E**). Direct and indirect immunoflorescence were consistent with PV. Anti- desmoglein 1 IgG (anti-Dsg1) and anti-desmoglein 3 IgG (anti-Dsg3) levels measured with ELISA were 1.8 and 13.6 U/mL respectively (normal <1.0 U/mL). Circulating autoantibodies against BP180 and BP230 were not detected. A final diagnosis of PV was made. Clinical remission was achieved through systemic corticosteroids and double-filtration plasmapheresis.

Case 2: A 28-year-old woman with a diagnosis of PV was treated in our clinic for 9 years. The patient had been clinically stable for years on low dose oral corticosteroid when she had a flare up during the 12th week of her first pregnancy. Unlike her previous clinical presentations, which were typical of mucocutaneous PV, extensive polycyclic plaques of fresh bullae encircling old crusted lesions appeared. These involved the trunk, extremities, palmoplantar areas, and scalp, accompanied by severe pruritus (**Figure 2A, B**). Mucosal areas were completely spared.

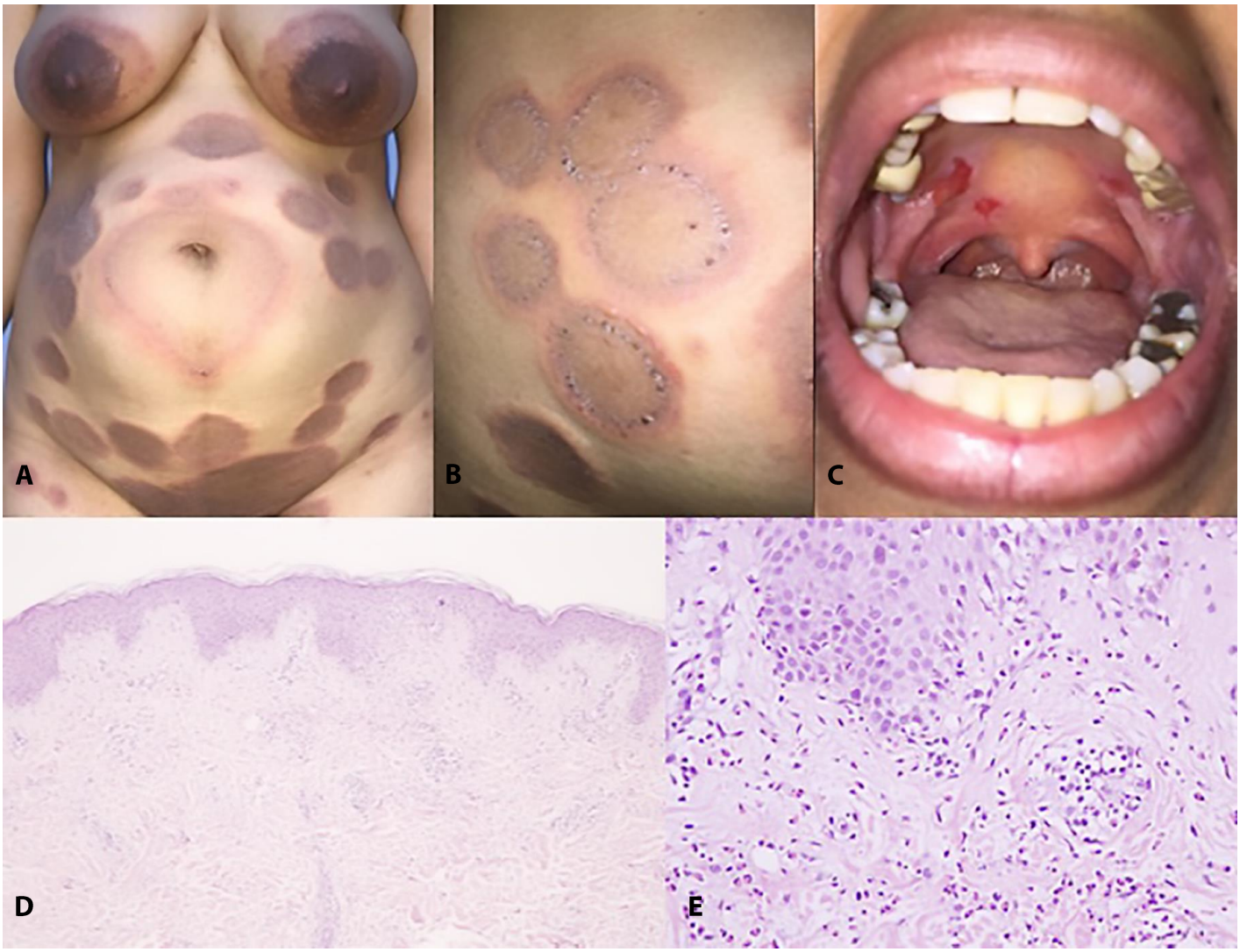

Figure 1. ***A-C)*** *Twenty-three-year-old female patient with severe erosions in the oral mucosa and annular blisters on the periphery of resolved lesions in a rosette like fashion involving the trunk and extremities on the 8th week of pregnancy.* ***D)*** *Although focal subepidermal detachment was suggestive of gestational pemphigoid, a diagnosis of PV was made with direct immunfluorescence and immunoserological findings. H&E, 100×.* ***E)*** *Eosinophilic spongiosis and marked eosinophilic infiltration in the papillary dermis can be seen. H&E, 400×.*

A biopsy revealed,acantholytic keratinocytes, edema in the papillary dermis, and an inflammatory perivascular infiltration containing large numbers of eosinophils extending to mid-dermis (**Figure 2C, D**). Intercellular deposition of IgG and C3 in the epidermis were seen by direct immunofluorescence microscopy. Indirect immunofluorescence microscopy performed on monkey esophagus also showed intercellular IgG deposition. Anti-Dsg1, and anti-Dsg3 levels, were measured as 5.8 and 2.0 U/mL, respectively. Despite the atypical clinical presentation the diagnosis of PV was confirmed. Systemic corticosteroid therapy was not sufficient so intravenous immune globulin (IVIG) treatment was initiated. Clinical remission was achieved after IVIG treatment (0.4g/kg/day for 5 consecutive days).

Case Discussion

The clinical features of the first patient resembled linear Ig A disease (LAD) and some subepidermal splitting was seen in histopathology. However, immunoserologic studies revealed the unexpected diagnosis of PV. Although the second patient already had a PV diagnosis, the peculiar clinical presentation prompted us to replicate the diagnostic procedures.

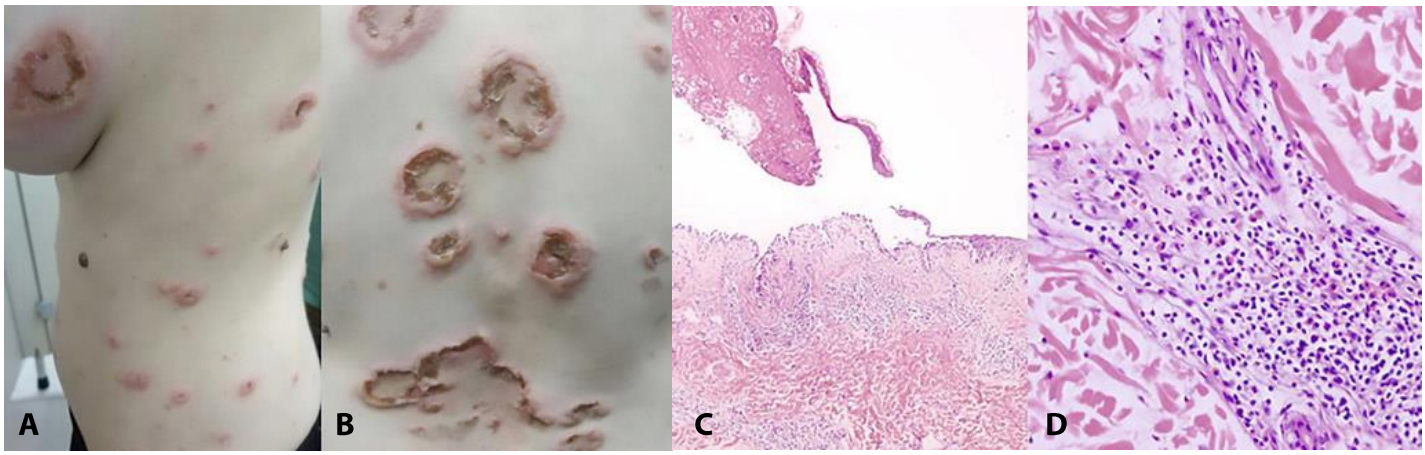

Figure 2. ***A, B****) Twenty-eight-year-old female with extensive polycyclic plaques of fresh bullae encircling old crusted lesions involving the trunk, extremities, palmoplantar areas and scalp, on the 12th week of her pregnancy.* ***C, D)*** *Suprabasal acantolysis accompanied by dense eosinophilic infiltration which becomes more prominent in the middle and deep portions of dermis. H&E,* ***C)****, 100×;* ***D)****, 400×.*

Both cases had a striking clinical appearance resembling LAD and to an extent, IgA pemphigus with blisters forming around resolving lesions in a rosette-like formation. Eosinophilic spongiosis and perivascular infiltration rich in eosinophils dominated the histopathology in both cases, which is an unusual finding for classical PV. It is worth noting that systemic corticosteroid therapy did not suffice and advanced treatment modalities were required in both patients. A case of cutaneous type PV similar to our second patient with positive anti-Dsg1 and anti-Dsg3 levels was reported [6]. Both patients gave birth without any complications and neonatal pemphigus was not seen.

Conclusion

To the best of our knowledge pregnancy associated pemphigus cases containing groups of bullae arranged in an annular fashion have not been reported in the literature. Changes in the immunologic and hormonal state during pregnancy may play a role in altering the classic clinical presentation and treatment response of PV.

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