Revisiting the "String of Pearls" Sign

Dear Editor,

The term "string of pearls" or "crown of jewels" appearance denotes the presence of multiple tense vesicles and bullae arranged circumferentially on an erythematous base. It has been characteristically described in linear IgA bullous dermatosis (LABD) in children and adults. Standard dermatology textbooks don't mention the association of this finding with other autoimmune bullous diseases.[1] Here, we describe two cases of bullous pemphigoid (BP) in adults which showed a string of pearl appearance.

Case 1: A 72-year-old female presented with a history of generalized itching and wheals followed by blistering for one month. Cutaneous examination revealed multiple tense bullae arranged in an annular pattern on the erythematous base, predominantly over the trunk and extremities.

Case 2: A 56-year-old male presented with a pruritic rash and blisters over his trunk and extremities for one month. Cutaneous examination revealed multiple erythematous annular plaques with tense bullae arranged peripherally, over the trunk and extremities [Figure 1]. Mucous membranes were spared in both.

Direct immunofluorescence (DIF) microscopy in both cases showed linear staining of the basement membrane zone (BMZ) with IgG and C3 [Figure 2]. Indirect immunofluorescence (IIF) on salt-split skin showed band localizing to the epidermal side of the split ('roof' pattern), thus confirming the diagnosis of BP [Figure 3].

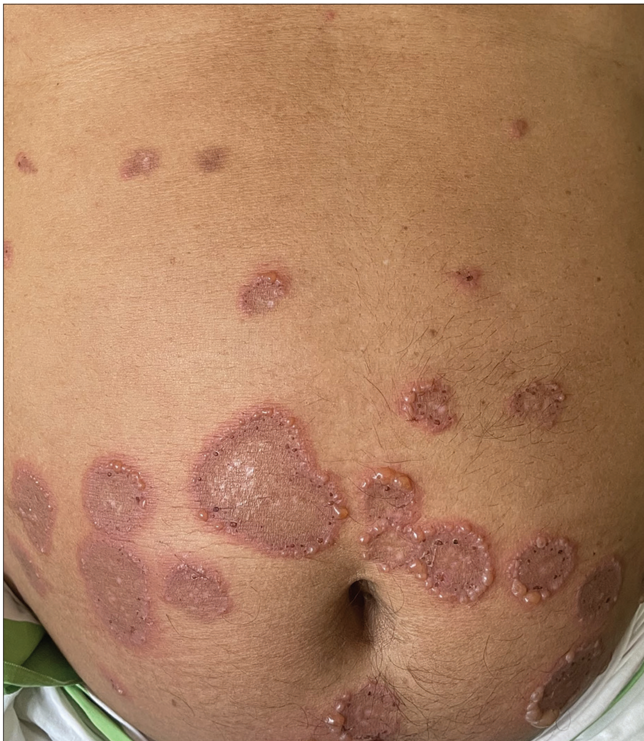


Figure 1: Multiple vesicles and bullae of varying sizes arranged in an annular pattern over erythematous base over the lower abdomen

Bullous pemphigoid (BP) is a subepidermal autoimmune bullous disorder (AIBD) that is usually seen in the elderly. It is characterized by the presence of tense blisters predominantly on the flexures and lower abdomen. Various atypical forms have been described in the literature; the presence of a 'string of pearl' or 'crown of jewel' appearance, a characteristic feature of LABD, has rarely been reported in association with BP.[1,2] Direct immunofluorescence (DIF) microscopy is considered the gold standard test in the diagnostic armamentarium of AIBD and helps to distinguish BP from LABD.

Linear staining of BMZ with IgA is characteristic of LABD, whilst in BP it is seen with IgG and C3.[3] Based on the clinical findings, a diagnosis of LABD was made; however, DIF and IIF microscopy helped us to confirm the diagnosis of BP.

Arès *et al*. have reported an unusual case of annular BP in a 50-year-old adult in whom a diagnosis of LABD and pemphigus herpetiformis was considered initially but finally, immunopathology helped to arrive at the correct diagnosis.[4] A similar string of pearls appearance has also been described in a case of infantile BP.[5] Subepidermal AIBDs exhibit overlapping clinical features and an attempt should be made in every case to arrive at the final diagnosis based on the DIF and serological tests. An accurate diagnosis is important in the management of these cases; while BP can be managed with topical or oral corticosteroids and tetracyclines, the treatment of choice for LABD is dapsone.[1]

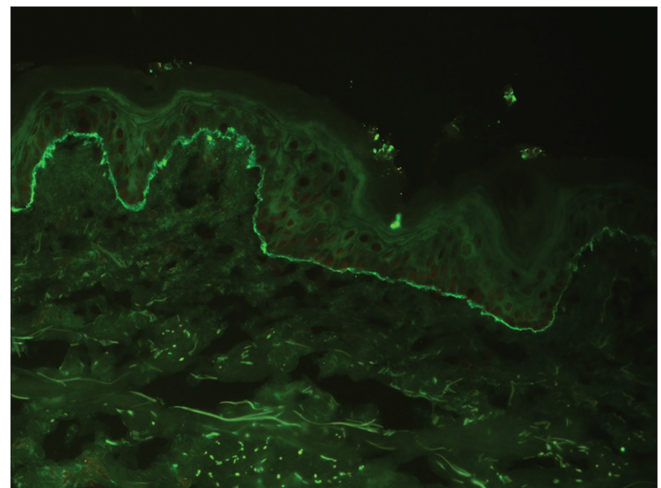


Figure 2: Direct immunofluorescence showing linear basement membrane zone staining with C3 (20x)

© 2023 Indian Dermatology Online Journal | Published by Wolters Kluwer - Medknow

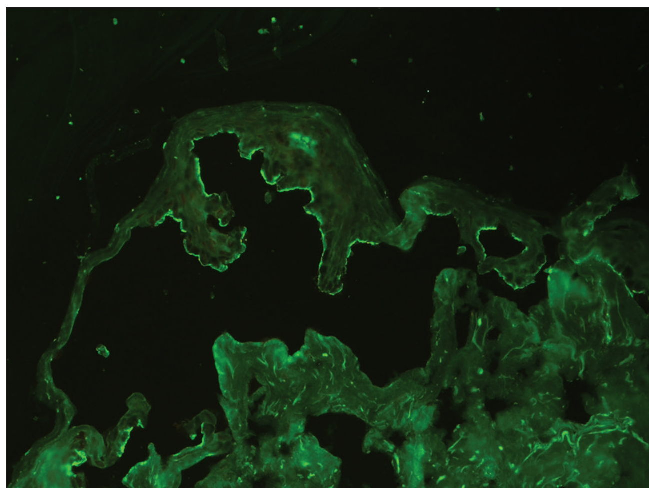


Figure 3: Salt split method showing linear basement membrane staining on epidermal side of split with IgG (20x)

To conclude, we would like to emphasize with a report that the “string of pearls” sign in adult AIBD is not characteristic of LABD and may occasionally be seen in BP. A final diagnosis of AIBD should not be made based on this sign alone and findings of immunological and serological tests should be taken into consideration. Knowledge of this unusual presentation will aid the clinician in the prompt diagnosis and initiation of appropriate treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Nivalika ASA, Krithiga Muralidharan[1], Raghavendra Rao

Department of Dermatology, Kasturba Medical College, Manipal, Manipal Academy of Higher Education, Manipal, Karnataka, [1]Meenakshi Medical College Hospital and Research Institute, Kanchipuram, Tamil Nadu, India

Address for correspondence:
Dr. Raghavendra Rao,
Professor and Head, Department of Dermatology, Kasturba Medical College, Manipal, Manipal Academy of Higher Education, Manipal, Karnataka - 576 104, Karnataka, India.
E-mail: raghavrao1@gmail.com

References

1. Schmidt E, Groves R. Immunobullous diseases. In: Griffiths CE, Barker J, Bleiker T, Chalmers R, Creamer D, editors. Rook’s Textbook of Dermatology. 9th ed. Oxford: Wiley Blackwell; 2016. p. 50.51-50.57.
2. Díaz-Morales P, Grau-Pérez M, De-La-Rosa-Del-Rey MP, Cárdenes-Santana MÁ. ‘String of pearls’ sign revealing linear IgA bullous disease. Am J Med 2021;134:e135-6.
3. Genovese G, Venegoni L, Fanoni D, Muratori S, Berti E, Marzano AV. Linear IgA bullous dermatosis in adults and children: A clinical and immunopathological study of 38 patients. Orphanet J Rare Dis 2019;14:115.
4. Arès S, Lim D, Jung S, Bouffard D, Mereniuk A. Annular bullous pemphigoid: A case report and review of literature. SAGE Open Med Case Rep 2022;10:2050313X221093128. Published 2022 Apr 21. doi: 10.1177/2050313X221093128.
5. Raposo I, Machado S, Sampaio R, Selores M. Infantile bullous pemphigoid with “string of pearls sign.” Dermatol Online J 2017; 23:13030/qt3fw4d2t2.

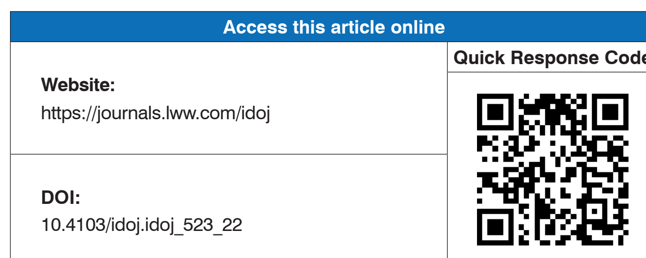
This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

Access this article online

Website: https://journals.lww.com/idoj

DOI: 10.4103/idoj.idoj_523_22

Quick Response Code



How to cite this article: Nivalika ASA, Muralidharan K. Rao R. Revisiting the “String of Pearls” sign. Indian Dermatol Online J 2023;14:567-8.

Received: 05-Oct-2022. **Revised:** 18-Dec-2022. **Accepted:** 26-Dec-2022. **Published:** 28-Jun-2023.

© 2023 Indian Dermatology Online Journal | Published by Wolters Kluwer - Medknow