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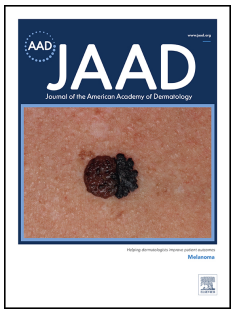

Brunsting-Perry pemphigoid: a retrospective case series of a frequently unrecognized condition

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PII: S0190-9622(20)32841-3

DOI: https://doi.org/10.1016/j.jaad.2020.10.029

Reference: YMJD 15327

To appear in: *Journal of the American Academy of Dermatology*

Received Date: 28 May 2020

Revised Date: 10 September 2020

Accepted Date: 13 October 2020

Please cite this article as: Imstepf V, Cazzaniga S, Beltraminelli H, Borradori L, Feldmeyer L, Brunsting-Perry pemphigoid: a retrospective case series of a frequently unrecognized condition, *Journal of the American Academy of Dermatology* (2020), doi: https://doi.org/10.1016/j.jaad.2020.10.029.

Brunsting-Perry pemphigoid: a retrospective case series of a frequently unrecognized condition

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word count text (excluding references, figures, and tables): 499
references count: 5
figure count: 1
table count: 1

Funding sources: None
IRB approval status: Reviewed and approved by the ethics committee of the Canton of Bern (KEK-2019-01811).
Conflicts of interest: The authors have no conflict of interest to declare.

Brunsting-Perry pemphigoid (BPP) constitutes a rare variant of mucous membrane pemphigoid (MMP). Since the first description of 7 cases, there have been only few case reports describing the characteristics of BPP.[1,2]

We have retrospectively assessed all BPP patients evaluated in our tertiary referral center between 2016 and 2019. The data was retrieved from our databank. Diagnosis of BPP was based on 1) clinical features 2) histopathological findings and 3) positive direct immunofluorescence microscopy studies with linear deposits of IgG, IgA and/or C3 along the dermo-epidermal junction.[3]

We identified 12 patients complying with the inclusion criteria. Patients' characteristics and findings are depicted in Table I. The mean age at presentation was 73 years, 80% of the patients were male; 36.4% had diabetes mellitus type II, 72.7% arterial hypertension, 54.5% chronic renal insufficiency and one suffered from colon cancer. There were three DPP4i-treated patients (25%), 4 patients (33.3%) received sartans. Ten of 11 patients (data missing for one patient) were treated with topical steroids. Among them, 8 had a relapse and required an additional treatment with methotrexate and/or tetracyclines. Complete remission, partial remission on minimal therapy and persistent disease were observed in 33.3%, 50%, and 16.7%, respectively.

This review of 12 cases represents the largest series of BPP reported so far in the English literature. Without any exceptions the lesions involved the head, face or neck, and in >40% also the upper body. More than 90% of our patients presented with erosions, ulcerations and prurigo-like lesions, while obvious blistering was absent in almost half the patients. (Fig.1) In one third of the cases, mild oral mucosal involvement occurred, which has been only anecdotally reported in BPP.[1,2] The polymorphic presentation likely explains the long mean diagnostic delay of 3 years observed in our series.

Immunologically, the majority of our BPP patients showed reactivity for BP180 and/or BP230, although the available data indicates that the targeted autoantigens of BPP are heterogenous, including type VII collagen and laminin-332.[2]

There is no validated therapeutic approach for BPP. Based on our observations, potent topical steroids should be the first-line therapeutic option. If the latter does not control the disease, either low dose methotrexate and/or tetracyclines appear to be

good second-line options.[3] These regimens resulted in either complete or partial control of the disease on minimal treatment in more than 80% of our cases. In our study, 7 BPP patients were either on DPP4is or sartans. Drug-induced MMP has been only anecdotally described.[2,4] DPP4is, as well as anti-PD1 inhibitors, have been associated with MMP and BP.[4] Hence, our findings suggest that DPP4is may be a potential trigger of BPP. Furthermore, sartans have been incriminated in triggering both BP and MMP.[4,5]

In summary, dermatologists should consider BPP in the presence of chronic localized erosions, ulcerations, blisters, and prurigo-like lesions with scarring, affecting the head and upper trunk in an elderly patient. Histologically, eosinophilia with erosion or subepidermal blistering is observed in the majority of the BPP cases. Nevertheless, the definite diagnosis depends on the results of the DIF microscopy findings.

Abbreviations

Brunsting-Perry pemphigoid (BPP)

mucous membrane pemphigoid (MMP)

direct immunofluorescence microscopy (DIF)

dipeptidyl peptidase-IV inhibitor (DPP4i)

programmed cell death protein-1 inhibitor (anti-PD1 inhibitors)

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Figure legend

Figure 1. Polymorphous clinical features of Brunsting-Perry pemphigoid: erythematous inflamed, light infiltrated area with scarring and milia formation in the preauricular and temporal region, respectively.

Table I. Demographics, clinical characteristics and histological parameters of the patients included

		N=12	%	Mean	SD
Gender	Female	2	16.7%		
	Male	10	83.3%		
Age at diagnosis (yrs)				73.1	11.2
Age at symptoms onset (yrs)				70.9	12.4
Diagnostic delay (yrs)				3.0	4.7
*Localization**		*12*			
Head/face/neck		12	100.0%		
Upper body		5	41.7%		
Oral		4	33.3%		
Other		1	8.3%		
*Clinical features**		*12*			
Bullous lesions		8	66.7%		
Ulcerations		11	91.7%		
Erosions		8	66.7%		
Scales		3	25.0%		
*Histological diagnosis**		*12*			
Spongiosis		1	8.3%		
Scarring/Fibrosis		5	41.7%		
Subepidermal blister		7	58.3%		
Eosinophils		8	66.7%		
Erosions		5	41.7%		
Other		3	25.0%		
*DIF**		*12*			
Linear C3/IgG		11	91.7%		
Other linear autoantibodies		5	41.7%		
Unspecific DIF		1	8.3%		
ELISA		*11*			
BP180		3	27.3%		
BP230		1	9.1 %		

BP180 + 230	1	9.1 %		
Negative	6	54.5%		
No data	1	-		
IIF	*10*			
Positive**	4	40.0%		
Negative	6	60.0%		
No data	2	-		
*ELISA + IIF**	*10*			
Both negative	4	40.0%		
At least one positive	6	60.0%		
Both positive	2	20.0%		
No data	2	-		

DIF: direct immunofluorescence, ELISA: enzyme-linked immunosorbent assay, IgG: immunoglobuline G, IIF: indirect immunofluorescence, yrs: years, N: number, SD: standard deviation

* More than one category may be present at the same time

** All IIF positive cases (4) were evaluated in the roof of the bulla