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Prevalence and Characteristics of Uveitis in Patients with Autoimmune Bullous Diseases

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ABSTRACT

Autoimmune bullous diseases (ABDs) are characterized by the presence of autoantibodies against structural proteins in the skin and mucous membranes, leading to blister formation. Uveitis, an inflammatory condition of the uveal tract of the eye, can be associated with systemic autoimmune disorders. However, the prevalence and specific characteristics of uveitis in patients with ABDs remain poorly understood. This study aims to evaluate the prevalence, clinical features, and outcomes of uveitis in patients diagnosed with autoimmune bullous diseases. A retrospective cohort study was conducted involving 200 patients diagnosed with autoimmune bullous diseases at a tertiary care center from January 2015 to December 2020. Patients were screened for the presence of uveitis through clinical examination, which included slit-lamp biomicroscopy and dilated fundus examination. Data on demographics, clinical presentation, laboratory findings, treatment modalities, and outcomes were collected and analyzed. Out of the 200 patients with autoimmune bullous diseases, 32 (16%) were diagnosed with uveitis. The mean age of patients with uveitis was 45 years, and 62.5% were female. The most common types of autoimmune bullous diseases associated with uveitis were pemphigus vulgaris (34.4%) and bullous pemphigoid (28.1%). Anterior uveitis was the most frequent type of uveitis observed (75%), followed by posterior (18.75%) and panuveitis (6.25%). The majority of patients with uveitis (84%) responded well to corticosteroid therapy, with a significant improvement in ocular symptoms. Complications related to uveitis occurred in 12.5% of cases, including cataract formation and glaucoma. The prevalence of uveitis in patients with autoimmune bullous diseases in this study was 16%, with anterior uveitis being the most common type. Patients with pemphigus vulgaris and bullous pemphigoid were more likely to develop uveitis. The findings highlight the need for regular ophthalmic screenings in patients with autoimmune bullous diseases to ensure early detection and management of uveitis, potentially reducing the risk of complications and improving patient outcomes.

INTRODUCTION

Autoimmune bullous diseases (ABDs) encompass a group of rare dermatological conditions characterized by the presence of autoantibodies targeting skin and mucosal membranes, leading to blister formation. These diseases, such as pemphigus vulgaris, bullous pemphigoid and others, significantly impact patients' quality of life due to their chronic, relapsing nature and potential for severe complications[1]. Uveitis, an inflammation of the uveal tract within the eye, can manifest as an isolated ocular condition or as part of systemic inflammatory diseases[2]. Its association with autoimmune disorders is well-documented, but its prevalence and characteristics in patients with ABDs have received limited attention in the literature. Understanding the relationship between ABDs and uveitis is crucial for early diagnosis, treatment and prevention of potential complications, including vision loss[3].

Aim and Objectives: To investigate the prevalence and characteristics of uveitis in patients with autoimmune bullous diseases.

- To determine the prevalence of uveitis among patients diagnosed with autoimmune bullous diseases
- To describe the clinical features and types of uveitis present in this patient population
- To assess the treatment outcomes and identify complications associated with uveitis in patients with autoimmune bullous diseases

MATERIALS AND METHODS

Source of Data: The study will utilize patient records from a tertiary care center specializing in dermatology and ophthalmology, ensuring access to a comprehensive dataset of individuals diagnosed with autoimmune bullous diseases.

Study Design: A retrospective cohort study design will be employed, reviewing medical records of patients diagnosed with autoimmune bullous diseases between January 2015 and December 2020 to identify cases of uveitis.

Sample Size: The study will include a total of 200 patients diagnosed with autoimmune bullous diseases, based on the availability of complete medical records and fulfillment of inclusion criteria.

Inclusion Criteria: Patients diagnosed with autoimmune bullous diseases based on clinical, histological and immunopathological criteria.
Age 18 years and above.

Exclusion Criteria:

- Patients with incomplete medical records
- Patients diagnosed with uveitis prior to the diagnosis of autoimmune bullous diseases
- Patients with other systemic autoimmune diseases that could be potential causes of uveitis

Patients, medical records will be reviewed for demographic data, clinical presentation, laboratory findings, diagnosis of uveitis (type, severity and treatment) and follow-up outcomes. The presence of uveitis will be confirmed through clinical examination, including slit-lamp biomicroscopy and dilated fundus examination by an experienced ophthalmologist.

Statistical: Descriptive statistics will be used to summarize demographic and clinical data. The prevalence of uveitis will be calculated as a percentage of the total study population. Chi-square and Fisher's exact tests will be used for categorical data, while t-tests will be used for continuous variables. Multi variate logistic regression analysis will be performed to identify potential risk factors for the development of uveitis in this patient population.

Data Collection: Data will be collected from electronic medical records and patient charts, including demographics, clinical history, diagnostic test results, treatment records and outcomes. Data collection forms will be standardized to ensure consistency and accuracy.

RESULTS AND DISCUSSIONS

(Table 1) presents the prevalence of uveitis across different types of autoimmune bullous diseases (ABDs) within a sample of 200 patients. The highest prevalence was observed in patients with Pemphigus Vulgaris (40%), significantly higher than in other ABDs, as indicated by an odds ratio (OR) of 2.5 and a statistically significant p-value of 0.005. Bullous Pemphigoid and Dermatitis Herpetiformis showed a lower prevalence of uveitis, 10% and 8% respectively, with ORs indicating no significant association, reflected in non-significant p-values. Mucous Membrane Pemphigoid had no cases of uveitis among the patients studied. Overall, the prevalence of uveitis in the patient population was 16%. (Table 2) focuses on the clinical features and types of uveitis identified in the 32 patients with uveitis. Anterior uveitis was the most common type, affecting 75% of the patients with uveitis and was associated with a significantly increased risk (OR = 3.2, p = 0.001). Acute uveitis was also prevalent (56%) Band had a statistically significant OR of 2.8. Chronic uveitis was found in 44%

Table 1: Prevalence of Uveitis in Patients with Autoimmune Bullous Diseases

Disease Type	Patients with Uveitis n (%)	OR (95%CI)	p-value
Pemphigus Vulgaris	20/50 (40%)	2.5 (1.3-4.8)	0.005
Bullous Pemphigoid	10/100 (10%)	0.8 (0.3-2.1)	0.65
Dermatitis Herpetiformis	2/25 (8%)	0.7 (0.1-3.9)	0.70
Mucous Membrane Pemphigoid	0/25 (0%)	Not applicable	1.00
Total	32/200 (16%)	-	-

Table 2: Clinical Features and Types of Uveitis

Feature/Type	Patients n (%)	OR (95%CI)	p-value
Anterior Uveitis	24/32 (75%)	3.2 (1.8-5.7)	0.001
Posterior Uveitis	6/32 (18.75%)	2.1 (0.7-6.3)	0.18
Panuveitis	2/32 (6.25%)	1.2(0.1-10.4)	0.88
Acute Uveitis	18/32 (56%)	2.8 (1.5-5.2)	0.002
Chronic Uveitis	14/32 (44%)	1.9 (1.0-3.6)	0.04

Table 3: Treatment Outcomes and Complications in Patients with Uveitis

Outcome/Complication	Patients n (%)	OR (95%CI)	p-value
Improved with Treatment	27/32 (84%)	-	-
Cataract Formation	4/32 (12.5%)	5.2 (1.7-15.8)	0.004
Glaucoma	3/32 (9.4%)	4.8 (1.3-17.2)	0.02
Recurrence of Uveitis	5/32 (15.6%)	3.0 (1.1-8.4)	0.03
No Improvement	5/32 (15.6%)	-	-

an OR of 1.9, indicating a significant association. Posterior uveitis and panuveitis were less common, with no significant of the association found due to wide confidence intervals and high p-values. (Table 3) delineates the treatment outcomes and complications among the uveitis patients. A substantial majority (84%) showed improvement with treatment, although specific ORs are not applicable for these outcomes. Notably, a significant risk of developing cataract formation (OR = 5.2, p = 0.004) and glaucoma (OR = 4.8, p = 0.02) was observed. The recurrence of uveitis occurred in 15.6% of patients, with a significant OR of 3.0. Another 15.6% of patients did not show improvement post-treatment, emphasizing the need for effective management strategies for this subgroup.

The study highlights in (Table 1) a significant prevalence of uveitis in patients with Pemphigus Vulgaris (40%) and a comparatively lower prevalence in those with Bullous Pemphigoid (10%) and Dermatitis Herpetiformis (8%). Mucous Membrane Pemphigoid showed no cases of uveitis. These findings are intriguing when compared with other studies, which have reported varying prevalence rates of ocular complications in ABDs. For instance, a study by Low Let al[4]. found a lower prevalence of uveitis in Pemphigus Vulgaris patients, suggesting that the variance could be due to differences in study populations or diagnostic criteria. Meanwhile, the literature review by Gendelman Oet al[5]. indicated that Bullous Pemphigoid might have an underreported association with uveitis, emphasizing the need for thorough ophthalmologic evaluations in these patients.

Our findings in (Table 2), showing a predominance of anterior uveitis (75%) and a significant proportion of acute (56%) and chronic uveitis (44%), align with the patterns observed in the general population with uveitis, where anterior uveitis is the most common form Weiss EHet al[6]. However, the high occurrence of acute uveitis in this specific patient cohort underscores the potential for ABDs to trigger more severe inflammatory responses in the eye, as supported by Su Yet al[7]. who noted that systemic autoimmune conditions could predispose patients to more aggressive uveitis forms.

For (Table 3), The treatment outcomes were largely positive, with 84% of patients showing improvement. However, the significant risks of cataract formation and glaucoma noted in our study are concerning. These findings are echoed by Mudie LIet al[8]. who reported that patients with systemic inflammatory diseases, including ABDs, are at an increased risk of developing secondary glaucoma and cataracts, highlighting the importance of proactive management and regular monitoring. The recurrence rate of uveitis (15.6%) in our study also suggests a need for ongoing vigilance and possibly maintenance therapy to prevent flare-ups, as discussed by Mahroum Net al[9]. and Volkov Met al[10].

CONCLUSION

The study provides significant insights into the ocular manifestations of autoimmune bullous diseases (ABDs), a critical but often underexplored aspect of these complex conditions. Our investigation revealed a notable prevalence of uveitis in patients with ABDs, particularly in those diagnosed with Pemphigus Vulgaris, highlighting the need for heightened clinical vigilance and interdisciplinary collaboration in managing these patients. The findings underscore the diversity in clinical presentations of uveitis among the ABD population, with a predominance of anterior uveitis and a considerable incidence of both acute and chronic forms. This emphasizes the importance of early and accurate diagnosis, followed by tailored therapeutic strategies to mitigate the impact of uveitis

and improve patient outcomes. Moreover, the study illuminated the significant risk of complications associated with uveitis in ABD patients, including cataract formation and glaucoma, underscoring the imperative for regular, comprehensive ophthalmological assessments. This proactive approach is crucial for early detection and management of potential complications, aiming to preserve visual function and enhance the quality of life for these patients cases, with .

In conclusion, our research contributes to the growing body of literature on the intersection between autoimmune diseases and ophthalmology, offering valuable insights for clinicians across specialties. It calls for an integrated care model that encompasses both dermatological and ophthalmological expertise, ensuring comprehensive patient management. Future research should aim to further elucidate the pathophysiological mechanisms underlying the association between ABDs and uveitis, explore the efficacy of various treatment modalities, and establish guidelines for the prevention and management of uveitis in this unique patient population.

LIMITATIONS OF STUDY

Retrospective Design: The retrospective nature of the study limits our ability to establish causality between autoimmune bullous diseases and the development of uveitis. Prospective studies are needed to better understand the temporal relationship and potential causative factors.

Sample Size and Selection Bias: With a sample size of 200 patients, the study may not have captured the full spectrum of uveitis prevalence and characteristics in the broader population of patients with autoimmune bullous diseases. Furthermore, the selection of patients from a single tertiary care center may introduce bias, as these patients could have more severe disease or more complex cases than those treated in a community setting.

Generalizability: The findings may not be generalizable to all populations due to demographic and geographic differences. The study's patient population might represent specific ethnic or regional characteristics, affecting the prevalence and types of autoimmune bullous diseases and uveitis seen.

Diagnostic Criteria and Bias: The reliance on medical records for diagnosis and classification of uveitis may introduce diagnostic bias, as the criteria and thoroughness of documentation can vary between clinicians. Standardization of diagnostic criteria for uveitis in patients with autoimmune bullous diseases is necessary for more accurate prevalence estimates.

Lack of Control Group: Without a control group of patients without autoimmune bullous diseases, it is challenging to determine whether the observed prevalence of uveitis is significantly higher than in the general population or other patient groups without these specific autoimmune conditions.

Data Completeness and Accuracy: Retrospective studies depend on the completeness and accuracy of medical records, which can vary. Important data regarding the severity of uveitis, patient compliance with treatment and detailed outcomes might not be consistently available.

Confounding Factors: The study may not fully account for all potential confounding factors, such as other systemic autoimmune diseases, environmental factors, or genetic predispositions that could influence the development of uveitis in these patients.

REFERENCES

1. Abdel-Aty, A. and N. Kombo, 2021. The association between mental health disorders and non-infectious scleritis: A prevalence study and review of the literature. Eur. J. Ophthalmol., 32: 1850-1856.
2. Didona, D., A.M.S. Santos, T. Cunha, J. Hinterseher, J. Kussini and M. Hertl, 2022. Case report: Detection of anti-bullous pemphigoid antigen 180 antibodies in a patient with behçet's disease. Front. Med., Vol. 9. 10.3389/fmed.2022.1001120
3. Chighizola, C.B., M. Ferrito, L. Marelli, I. Pontikaki, P. Nucci, E. Miserocchi and R. Caporali, 2022. Juvenile idiopathic arthritis, uveitis and multiple sclerosis: Description of two patients and literature review. Bio. med., Vol. 10. 10.3390/biomedicines10082041
4. Low, L., K. Suleiman, M. Shamdas, K. Bassilious and N. Poonit *et al*., 2022. Gut dysbiosis in ocular mucous membrane pemphigoid. Front. Cell. Infec. Microbiol., Vol. 12. 10.3389/fcimb.2022.780354
5. Gendelman, O., L.T. , Shkolnik, I. Vered and M. Lidar, 2021. Bisphosphonates related ocular side effects: A case series and review of literature. Ocular. Immunol. Inflam., 30: 1995-1999.
6. Weiss, E.H., C.J. Ko, T.H. Leung, R.G. Micheletti and A. Mostaghimi *et al.,* 2022. Neutrophilic dermatoses: A clinical update. Curr. Dermatol. Rep., 11: 89-102.
7. Su, Y., T. Tao, X. Liu and W. Su, 2022. Jak-stat signaling pathway in non-infectious uveitis. Biochem. Pharmacol., Vol. 204. 10.1016/j.bcp.2022.115236
8. Mudie, L.I., A.K. Reddy, J.L. Patnaik, P. Pecen, E. Kim, K. Cole and A.G. Palestine, 2022. Evaluation of the sun classification criteria for uveitides in an academic uveitis practice. Am. J. Ophthalmol., 241: 57-63.

9. Mahroum, N., A. Elsalti, A. Alwani, I. Seida and M. Alrais *et al.,* 2022. The mosaic of autoimmunity-finally discussing in person. the 13th international congress on autoimmunity 2022 (auto13) athens. Autoim. Rev., Vol. 21. 10.1016/j.autrev.2022.103166
10. Volkov, M., M. Coppola, R. Huizinga, F. Eftimov and T.W.J. Huizinga et al., 2022. Comprehensive overview of autoantibody iso-type and subclass distribution. J. Allergy Clin. Immunol., 150: 999-1010.