

evidenciaron elevaciones de la PCR, la beta2 microglobulina y el complemento CH50, revelando un estado inflamatorio.

La variante p.(Tyr135His) no se encuentra descrita en las bases de datos clínicas ni en la bibliografía científica consultada hasta el momento de realizar este trabajo. Sí aparece anotada en la base de datos dbSNP (*database of Single Nucleotide Polymorphisms*) (rs145078602), pero no en la base de datos de frecuencia poblacional gnomAD (*The Genome Aggregation Database*). El predictor bioinformático CADD (*Combined Annotation Dependent Depletion*) estima que el cambio tendría un efecto tolerado (< 10).

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The Latin-American rheumatology community needs to put the eye on ocular cicatricial pemphigoid



La comunidad reumatológica latinoamericana necesita poner el ojo en el penfigoide cicatricial ocular

Dear Editor,

Mucous membrane pemphigoid is a systemic and scarring autoimmune disease. Isolated ocular involvement has classically been called ocular cicatricial pemphigoid (OCP). This entity can lead to irreversible blindness if not treated early.^{1,2} Treatment is always systemic; thereby, ophthalmologists seek the support of rheumatologists, dermatologists, or a clinician with autoimmunity knowledge.³ Given the lack of epidemiological data in our region, the seriousness of this pathology, and it is supposed unfamiliarity by medical professionals of these specialties, it is proposed to carry out this work. Our objective was to know the familiarity of rheumatologists and ophthalmologists with OCP in our region.

A survey was conducted online through a Google form link. The questionnaire was primarily directed toward rheumatologists and ophthalmologists who are members of scientific societies in America. The questionnaire consisted of 13 questions about the age, sex, country, specialty, principal place of work (public or private institution or private practice), whether or not they are familiar with the entity, how many patients they see annually, the specialties with which they work to address these patients, diagnostic methodology (clinical vs. biopsy), the treatments used, and the difficulties in approaching these patients in daily practice. For those physicians who saw at least two patients with OCP in the last year and answered that they were familiar, we considered them familiar and up-to-date with the disease.

We received 463 surveys, and 433 were included, excluding duplications and those surveyed that did not belong to an Ame-

rican country. Three hundred-seven (70.9%) were rheumatologists, and 112 (25.9%) were ophthalmologists. Other specialties were dermatologists, immunologists, and clinicians. Most participants were from Argentina, Colombia, Chile, and Mexico. **Table 1** describes the general characteristics of the respondents.

Forty percent reported being familiar with the OCP. Among them, 66.1% were ophthalmologists and 31.3% were rheumatologists. When we analyzed only the group of ophthalmologists' familiarity with the OCP, we observed that the specialty to which these patients are most referred is rheumatologists (85%).

The main difficulties reported in the management of these patients were: (1) the lack of information, and diffusion of the OCP in the different scientific activities and texts of the specialties involved in treating this entity; (2) the difficulty in accessing and performing the biopsy of the ocular conjunctiva; (3) the difficulties in communication between specialists to carry out an appropriate follow-up.

Like other authors,^{2–4} we emphasize the key to success with this entity is to have an early multidisciplinary approach. Considering the main difficulties, we believe that more focus should be placed on increasing the scientific activities related to this disease in congresses and journals, mainly related to rheumatology. Furthermore, it is important to address the challenges associated with biopsies that are likely caused by factors such as the high costs of immunofluorescence, inadequate insurance coverage, and the lack of pathologists specialized in OCP.⁴

A limitation of this study is that the survey distribution may have been different per country. Also, the low number of dermatologists involved maybe was that the survey was primarily driven by the rheumatology field, which may not have effectively reached out to many dermatologists.

Observing the high percentage of unfamiliarity with this entity by rheumatologists (70%) and the significant referral of these patients to rheumatology by ophthalmologists familiar with OCP

Table 1

General characteristics of participants by whether or not they are familiar with ocular cicatricial pemphigoid.

	Total n = 433	Familiar with OCP n = 173 (39.9%)	Unfamiliar with OCP n = 260 (60.1)	p value
<i>Females, n (%)</i>	252 (58.6)	92 (36.5)	160 (63.4)	0.06
<i>Age (years), mean (SD)</i>	46.6 (11.6)	47.8 (11.2)	45.8 (11.8)	0.08
<i>Specialties</i>				
Rheumatologists, n (%)	307 (70.9)	96 (31.3)	211 (68.7)	<0.001
Ophthalmologists, n (%)	112 (25.9)	74 (66.1)	38 (33.9)	<0.001
Other specialties, n (%)	14 (3.2)	3 (1.7)	11 (4.2)	0.17
<i>Professional practice</i>				
Public institutions, n (%)	231 (53.3)	79 (45.6)	152 (58.5)	0.01
Private institutions, n (%)	269 (62.1)	121 (69.9)	148 (56.9)	0.01
Private practice, n (%)	203 (46.9)	85 (49.1)	118 (45.4)	0.44
<i>Patients treated with OCP in the last year</i>				
Between 0 and 1, n (%)	23 (5.4)	0	23 (8.8)	<0.001
Between 2 and 10, n (%)	379 (88.7)	147 (84.9)	232 (89.2)	0.18
More than 10, n (%)	25 (5.8)	24 (13.8)	1 (0.4)	<0.001
<i>Used systemic drugs</i>				
Methotrexate, n (%)	220 (50.8)	133 (76.9)	87 (33.5)	<0.001
Azathioprine, n (%)	113 (26.1)	68 (39.3)	45 (17.3)	<0.001
Mycophenolate, n (%)	72 (16.6)	51 (29.5)	21 (8.1)	<0.001
Cyclophosphamide, n (%)	55 (12.7)	39 (22.5)	16 (6.1)	<0.001
Rituximab, n (%)	63 (14.5)	46 (26.5)	17 (6.5)	<0.001
Dapsone, n (%)	33 (7.6)	27 (15.6)	6 (2.3)	<0.001
Immune globulin therapy, n (%)	11 (2.5)	10 (5.7)	1 (0.4)	0.001
Anti-TNF, n (%)	31 (7.2)	21 (12.1)	10 (3.8)	0.001
<i>Diagnosis by biopsy, n (%)</i>	196 (45.5)	121 (69.9)	75 (28.8)	<0.001

OCP: ocular cicatricial pemphigoid; n: number; SD: standard deviation; TNF: tumor necrosis factor- α .

(85%), it is imperative to work to expand the knowledge, and joint effort between both specialties, to improve and standardize the approach to this entity, which produces irreversible sequelae on the ocular surface.

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