Anterior segment parameters associated with extramuscular manifestations in polymyositis and dermatomyositis

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Abstract

- **AIM:** To evaluate detailed anterior segment parameters of patients with idiopathic inflammatory myopathies (IIM), including polymyositis (PM), and dermatomyositis (DM), and to clarify the associations between these data and clinical variables of IIM.
- **METHODS:** Totally 57 PM, 41 DM patients and 62 controls were enrolled in this cross-sectional, observational, case-control study. All study participants underwent Pentacam evaluation. Laboratory investigations consisted of different antibody assays, while extramuscular clinical assessments included Raynaud’s phenomenon, dysphagia, interstitial lung disease, arthritis/arthralgia, and weight loss. Objective signs and subjective symptoms of dry eye disease (DED) were also evaluated.

**RESULTS:** All pachymetric parameters [center, apex, thinnest and maximal keratometry (K max)] and corneal volume (CV) of both sides of PM patients proved to be significantly lower. Some pachymetric data were also noticed as significantly decreased compared to those of controls. Several significant differences were traced between anterior segment values and extramuscular manifestations of myositis, largely in case of arthritis/arthralgia and weight loss, whereas associations between anterior segment parameters and antibodies were weak. Objective clinical tests of DED were also significantly decreased in IIM patients.

**CONCLUSION:** The results suggest that all IIM patients have thinner corneas compared with those of controls, and decreased corneal parameters are significantly associated with the occurrence of some extramuscular manifestations. In addition, IIM patients tend to develop objective signs of DED.

**KEYWORDS:** dry eye; extramuscular manifestations; dermatomyositis; polymyositis; Scheimpflug imaging

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INTRODUCTION

The group of idiopathic inflammatory myopathies (IIM), also accepted as autoimmune myositis, represents diverse, systemic rheumatic diseases identified by chronic weakness and inflammatory cell infiltrates in the skeletal muscles[1]. IIM incorporate various subgroups, including polymyositis (PM), dermatomyositis (DM), inclusion body myositis (IBM), and amyopathic dermatomyositis (ADM) in adults, and juvenile dermatomyositis (JDM) in children, while three other subgroups of small sample sizes have also been classified: juvenile PM, immune-mediated necrotizing myopathy (IMNM), and hypomyopathic DM[2]. IIM affects multiple organs and systems, including principally the...
muscles, but also often the skin, joints, and lungs, and even
the eyes. Ocular involvement of IIM encompasses a broad
spectrum of complications of adjacent structures and diverse
anterior and posterior segment damage to the eyes. There is
only scant evidence published in the literature regarding ocular
manifestations of IIM and most of them come from limited
case studies and reports. Previously corneal changes in IIM
have been extenuated despite the fact that cornea is especially
predisposed to intra-and postoperative complications during
corneal laser interventions, cataract surgeries and keratoplasties
in autoimmune diseases.[3-5]. Recently ophthalmic investigations
in connection with immune-related and connective tissue
disorders have become more prominent since ocular
impairments can be traced as part of various systemic immune-
based disorders.

As far as is known, there has been no other, but our own
previous study that dealt with corneal involvement of PM
and DM patients[6], but no investigation of detailed anterior
segment values of the eye has been presented so far in
patients with IIM, consequently our primary aim was to
obtain comprehensive data about anterior segment of IIM
patients with Scheimplug imaging. Our secondary aim was
to elucidate relationship between these quantities and clinical
parameters of IIM.

SUBJECTS AND METHODS

Ethical Approval The study protocol was approved by the
local ethics committee (HBR/052/00204/2014; DE OEC
RKEB/IKEB 4071-2013) and was in full compliance with
Good Clinical Practices (GCP) guidelines of the European
Union, and the Declaration of Helsinki. Informed consent was
collected from all patients and controls.

Characteristics of Patients Consecutive PM and DM patients
diagnosed according to the corresponding international
criteria[7] were selected into the study. All patients visited the
Department of Immunology of the University of Debrecen, and
none of them suffered from secondary Sjögren’s syndrome,
because all of them has normal ranged anti-Sjögren’s syndrome
related antigen A, and B (SS-A and SS-B) test values. Ocular
examinations, containing noncycloplegic best spectacle-
corrected visual acuity (BCVA) in Snellen’s chart, a regular
slit lamp investigation in order to exclude any irregularities of
the ocular surface, tear film or adnexa, and intraocular pressure
determination, were also performed for all contributors.

Features of Controls Controls were recruited out of
the participants who entered the Department of Ophthalmology
of the University of Debrecen for routine ophthalmological
check-ups, and had minor refractive errors (±1.0 diopter),
without any known systemic diseases such as diabetes mellitus,
Sjögren’s syndrome, rheumatic diseases, connective tissue
diseases (Sharp syndrome, mixed connective-tissue disease,
undifferentiated connective tissue disease, Ehlers-Danlos
syndrome, epidermolysis bullosa, etc.) at the same time. The
same ophthalmological examinations were carried out as in
patients.

Exclusion Criteria Eye drop administration within two
weeks preceding examinations was an exclusion criterion for
all participants, and no eye drops were allowed during the
study period. Regarding further exclusion criteria irregular
eyelid position and closure, contact lens wearing, history of
ocular surgery, presence of ocular infection, inflammation of
cslera, episcleral layer or uvea, trauma of the eye, corneal haze,
peripheral or central corneal melting.

Ophthalmological and Clinical Tests To examine dry eye
disease (DED) both tear break-up time (tBUT) and Schirmer-I
tests (STI) were used to analyze tear film stability and tear
production. For the former a strip of fluorescein (Haag-Streit,
Koenitz, Switzerland) wetted with a drop of unpreserved,
sterile saline solution 0.9% was used. This strip was touched
to the lower part of the bulbar conjunctiva causing minimal
stimulus and followed by some blinking the tear film was
inspected under cobalt blue light of a slit lamp. The tBUT
value was determined as the interval between the final entire
blink and the first appearance of a dry spot and was given in
seconds. Three evaluations were done in each eye of every
subject and the average of the three quantities was considered
as mean value.

To investigate tear production un-anesthetized Schirmer
test, i.e. the STI was used. In the course of this procedure
standardized strips of filter paper (Alcon Laboratory, Fort
Worth, Texas, USA) were placed at the lower-lid margin at
the junction of the middle and temporal third of each eye, and
during the maneuver special care was taken not to touch the
cornea. Participants were requested to avoid eye and eyelid
movements for 5min followed by a careful eye closure, next
the strip was extracted. STI was determined by measuring the
wet part of the strip (mm/5min). Regarding the subjective
symptoms of DED the Ocular Surface Disease Index (OSDI)
questionnaire (provided by Allergan, Inc., Irvine, CA, USA)[8]
was adopted since this is highly recommended by the U.S. Food
and Drug Administration (FDA) for use in clinical studies[9].
As for IOP quantification Huvitz HNT-1P (Huvitz, Dongan-
gu, Republic of Korea) noncontact tonometer was applied, and
three evaluations per eye were performed and the average of
them was determined as the mean value. Furthermore, ages at
the time of diagnosis, disease duration of patients were also
taken into consideration. Regarding laboratory quantities anti-
histidyl-tRNA synthetase antibody (Jo-1), anti-nuclear factor
(ANF), beta-2-glycoprotein (β2GPI), both anti-Sjögren’s-
syndrome-related antigen A (SS-A), and B (SS-B), metaphase
chromosomes, anti-cyclic citrullinated peptide (CCP), and
out of specific antigens anti-transcription intermediary factor 1-gamma (TIF1-γ), anti-Mi-2, anti-Pm/Scl (polymyositis/scleroderma), and extractable nuclear antigen (ENA) were measured. As for clinical parameters the assessment of Raynaud’s phenomenon, dysphagia, mechanic’s hands, interstitial lung disease (ILD), arthritis/arthralgia, muscle pain, and weight loss were examined.

**Pentacam Examinations** Regarding anterior segment evaluations a Pentacam (Pentacam HR, Oculus Optikgeräte GmbH, Wetzlar, Germany) was applied and undilated pupils were used in order to avoid fluorescein dying. Participants were requested to fix their chin on the chinrest while their foreheads touched the headband. Corneal power of both flat (K1) and steep (K2) axes, also mean corneal power (Km), and pachymetric measurements [pachy center, pachy apex, thinnest location and maximal keratometry (Kmax)] were noted. In addition, corneal volume (CV), anterior chamber volume (ACV), and anterior chamber depth (ACD), anterior chamber angle width (ACA) were determined besides pupil size. The averages of three consecutive quantifications were accepted for the study.

To exclude the issue of diurnal alterations all classifications were completed between 9:00 a.m. and 11:00 a.m. All measurements were performed on successive days, and constant temperature, light, and humidity conditions were applied to eliminate any ocular surface stress.

**Statistical Analyses** Continuous variables were characterized as the mean with standard deviation (SD), while categorical variables were defined as frequencies and their percentages.

The distribution of the data was monitored by applying the Kolmogorov-Smirnov test, while Mann-Whitney U test was utilized assuming nonparametric distribution. To compare categorical data, Chi-square test and Fisher’s exact test were applied. Correlation coefficients between variables were counted using Spearman’s method. P values less than 0.05 were considered significant. For the statistical analysis IBM SPSS 25 statistical software (GraphPad Software Inc., San Diego, CA, USA) was employed.

**RESULTS**

**Characteristics of Patients and Controls** A total of 57 patients with PM (45 females and 12 males), mean age 57.70±9.19y and 41 patients with DM (26 female and 15 male, mean age 45.39±11.23y, were recruited into our study. The mean disease duration was 12.74±9.91 and 10.20±7.37y, respectively. None of the patients received immunomodulatory therapy, including cyclosporine A (CsA). During examinations, the basic disease was inactive, only a maintenance treatment with methylprednisolone (4 mg/die) was used.

Totally 62 gender-, age-, and refraction-matched controls (50 females and 12 males, mean age 58.32±9.41y were also enrolled. All participants were of Caucasian origin. No statistically significant differences in age and gender between the two groups were detected. Data are presented in Table 1.

**Outcome of Ocular Examinations** Regarding subjective symptoms of DED the average OSDI score was 23.00±21.54 for PM patients, 15.62±11.83 for DM patients, and 14.94±9.60 for control subjects. There was no significant difference in OSDI score values between different groups.
Objective signs of DED both Schirmer-I and tBUT test results were found to be significantly lower as compared to those of healthy controls. No significant difference was found in BCVA and IOP values between IIM patients and healthy volunteers. Data are also represented in Table 1.

**Pentacam Data** Regarding Pentacam data, significantly lower parameters were detected between each pachymetric measurement (center, apex, thinnest, Kmax) and CV values for PM patients in both sides and some pachymetric data were also traced to be significantly reduced compared to those of the control group, moreover, except for Kmax differences of pachymetric quantities between PM and DM patients were significant (Table 2).

**Outcome of Clinical and Laboratory Examinations** Raynaud’s phenomenon occurred in 28 (49%) of 57 PM and 15 (37%) of 41 DM patients, mechanic’s hand was found in 7 (12%) and 6 (15%), dysphagia was present in 7 (12%) and 19 (46%), ILD developed in 18 (32%) and 14 (34%), arthritis/arthralgia in 35 (61%) and 22 (54%), muscle pain in 32 (56%) and 25 (61%), and weight loss in 3 (5%) and 5 (12%) patients, respectively. Regarding occurrence of extramuscular manifestations only dysphagia was represented significantly in DM patients, as shown in Table 3. As for antibodies, ANF occurred in 21 PM patients and 22 DM patients, β2GPI in 7 and 4, anti-CCP in 3 and also 3, anti-Jo-1 in 10 and 3, ENA in 6 and 1, metaphase chromosomes in 9 and 10, and anti-Pm/Scl in 2 and 3 patients, respectively.

Table 4 shows the results of the associations between anterior segment data and antibodies when patients were investigated together. In general, the associations were weak, with only sporadic significant differences traced. However, numerous significant differences were traced between anterior segment values and extramuscular manifestations of myositis, largely in case of arthritis/arthralgia and weight loss; data are given in Table 5. Concerning correlation between Pentacam data and age or disease duration of IIM patients a significant negative

**Table 2 Anterior segment data of PM and DM patients and controls**

<table>
<thead>
<tr>
<th>Parameters</th>
<th>PM (n=57)</th>
<th>DM (n=41)</th>
<th>Control (n=62)</th>
<th>P (PM vs control)</th>
<th>P (DM vs control)</th>
<th>P (PM vs DM)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pachy center (µm)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>548.18±28.50</td>
<td>558.37±27.54</td>
<td>570.94±28.37</td>
<td>0.001</td>
<td>0.039</td>
<td>0.038</td>
</tr>
<tr>
<td>L</td>
<td>550.02±29.91</td>
<td>561.24±26.46</td>
<td>571.58±26.49</td>
<td>0.002</td>
<td>0.116</td>
<td>0.034</td>
</tr>
<tr>
<td>Pachy apex (µm)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>550.65±29.30</td>
<td>560.44±27.27</td>
<td>556.58±97.29</td>
<td>0.005</td>
<td>0.094</td>
<td>0.049</td>
</tr>
<tr>
<td>L</td>
<td>552.00±30.59</td>
<td>563.10±26.48</td>
<td>572.71±27.20</td>
<td>0.003</td>
<td>0.174</td>
<td>0.028</td>
</tr>
<tr>
<td>Pachy thinnest (µm)</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>543.00±29.08</td>
<td>552.17±26.45</td>
<td>566.35±28.15</td>
<td>0.001</td>
<td>0.019</td>
<td>0.046</td>
</tr>
<tr>
<td>L</td>
<td>544.63±30.62</td>
<td>554.63±27.00</td>
<td>567.68±26.93</td>
<td>0.001</td>
<td>0.065</td>
<td>0.044</td>
</tr>
<tr>
<td>Kmax (D)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>45.39±2.14</td>
<td>44.55±1.78</td>
<td>43.96±1.16</td>
<td>&lt;0.001</td>
<td>0.195</td>
<td>0.051</td>
</tr>
<tr>
<td>L</td>
<td>45.28±1.70</td>
<td>44.68±1.71</td>
<td>44.04±1.14</td>
<td>&lt;0.001</td>
<td>0.107</td>
<td>0.105</td>
</tr>
<tr>
<td>CV (mm³)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>59.70±2.83</td>
<td>60.31±3.15</td>
<td>61.68±3.57</td>
<td>0.006</td>
<td>0.084</td>
<td>0.187</td>
</tr>
<tr>
<td>L</td>
<td>60.12±3.39</td>
<td>61.12±4.38</td>
<td>61.78±3.13</td>
<td>0.022</td>
<td>0.172</td>
<td>0.306</td>
</tr>
<tr>
<td>ACV (mm³)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>132.26±32.10</td>
<td>137.22±32.87</td>
<td>135.42±28.39</td>
<td>0.429</td>
<td>0.842</td>
<td>0.336</td>
</tr>
<tr>
<td>L</td>
<td>134.54±34.22</td>
<td>140.15±33.57</td>
<td>140.03±28.38</td>
<td>0.378</td>
<td>0.982</td>
<td>0.347</td>
</tr>
<tr>
<td>ACD (mm)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>4.05±5.63</td>
<td>3.46±0.73</td>
<td>3.31±0.49</td>
<td>0.339</td>
<td>0.716</td>
<td>0.306</td>
</tr>
<tr>
<td>L</td>
<td>3.41±0.73</td>
<td>3.41±0.66</td>
<td>3.37±0.52</td>
<td>0.510</td>
<td>0.941</td>
<td>0.562</td>
</tr>
<tr>
<td>KPD</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>1.23±0.13</td>
<td>1.17±0.19</td>
<td>1.22±0.15</td>
<td>0.678</td>
<td>0.279</td>
<td>0.078</td>
</tr>
<tr>
<td>L</td>
<td>1.26±0.19</td>
<td>1.18±0.15</td>
<td>1.22±0.15</td>
<td>0.386</td>
<td>0.443</td>
<td>0.082</td>
</tr>
<tr>
<td>ACA (°)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R</td>
<td>32.74±7.39</td>
<td>34.85±7.73</td>
<td>31.84±6.61</td>
<td>0.892</td>
<td>0.088</td>
<td>0.107</td>
</tr>
<tr>
<td>L</td>
<td>33.91±8.12</td>
<td>34.34±7.88</td>
<td>32.95±6.12</td>
<td>0.865</td>
<td>0.528</td>
<td>0.687</td>
</tr>
</tbody>
</table>

PM: Polymyositis; DM: Dermatomyositis; Kmax: Maximal keratometry; CV: Corneal volume; ACV: Anterior chamber volume; ACD: Anterior chamber depth; KPD: Keratometric power deviation; ACA: Anterior chamber angle width.
correlation was only found between ACV and age of patients, while a significant positive correlation was observed between \( K_{\text{max}} \) and disease duration. Data are presented in Figures 1 and 2.

**DISCUSSION**

IIM is an uncommon acquired, systemic disease, the etiopathogenesis of which is unknown as yet, but genes within the major histocompatibility complex (MHC), the most significant genetic risk factor and the interferon pathway have been identified in its formation\(^{[10]}\). Incidence rates range from 1.16 to 19/1000 000 per year with prevalence rates between 2.4 and 33.8/100 000 inhabitants\(^{[11]}\). There is only an insufficient number of papers regarding ocular involvement in IIM, principally manifestations of the adnexa or posterior segment changes. Heliotrope rash, a characteristic skin finding in DM has been the most widely discussed finding\(^{[12-15]}\). In addition, diverse unlimited inflammations in the anterior segment, such as conjunctivitis, iritis, and episcleritis have also been described\(^{[16-17]}\). In a study carried out among patients with rheumatic diseases DED was also delineated in PM patients\(^{[18]}\). Regarding posterior segment of the eye, DM related retinopathy was first described by Bruce\(^{[19]}\) in 1938, followed by several case reports later on\(^{[20-24]}\), while Purtscher-like retinopathies in DM were depicted not long ago\(^{[25-26]}\). In a survey asymptomatic retinopathy was noticed in 14% of patients with PM and DM\(^{[27]}\). Lowder et al\(^{[28]}\) described a 49-year-old Japanese DM patient presenting initially with unilateral retinal pigment epithelial (RPE) detachment, and later with the fellow eye’s serous retinal detachment (SRD) at the macular area as well. Furthermore, his visual acuities and SRD fluctuated with exacerbations and remissions of the underlying disease. Ocular myositis associated with DM is a rare phenomenon in the course of the disease and two publications point out that it might be missed by clinicians\(^{[29-30]}\). Based on the abovementioned papers, the eye is expected to be involved in IIM. However, anterior segment lesions mainly corneal impairments have been unfairly put into the shade even if corneal structure and function considerably determined by highly ordered hierarchical organization of collagen fibrils can vary in immune-related and connective tissue diseases\(^{[31]}\). To date, there is still no study evaluating anterior segment findings in detail in IIM patients and their associations with clinical parameters have not been investigated either. The assessment of anterior segment values including corneal quantities is of vital importance for various diagnostic and therapeutic approaches and accordingly different modern imaging technologies have been developed. In the present study accurate anterior segment values of PM and DM patients were quantified, and additionally, objective signs and subjective symptoms of DED were also evaluated, since DED and corneal involvement are usual ocular findings in patients with rheumatic diseases\(^{[32-34]}\).

Based on our investigations PM patients rather than DM tend to develop thinner and lower volume corneas, since in terms of Pentacam data the former has more impaired corneal values. Concerning DED both PM and DM patients were found to have significantly reduced measured objective clinical test

<table>
<thead>
<tr>
<th>Table 3 Extramuscular findings in PM and DM patients</th>
</tr>
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<tbody>
<tr>
<td>Parameters</td>
</tr>
<tr>
<td>Raynaud +</td>
</tr>
<tr>
<td>Raynaud -</td>
</tr>
<tr>
<td>Mechanic’s hands +</td>
</tr>
<tr>
<td>Mechanic’s hands -</td>
</tr>
<tr>
<td>Dysphagia +</td>
</tr>
<tr>
<td>Dysphagia -</td>
</tr>
<tr>
<td>ILD +</td>
</tr>
<tr>
<td>ILD -</td>
</tr>
<tr>
<td>Arthritis/arthritis +</td>
</tr>
<tr>
<td>Arthritis/arthritis -</td>
</tr>
<tr>
<td>Muscle pain +</td>
</tr>
<tr>
<td>Muscle pain -</td>
</tr>
<tr>
<td>Weight loss +</td>
</tr>
<tr>
<td>Weight loss -</td>
</tr>
</tbody>
</table>

ILD: Interstitial lung disease.
### Table 4: Association between anterior segment data and different antibodies of IIM patients

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Jo-1</th>
<th>ANF</th>
<th>β2GPI</th>
<th>Metaphase chromosomes</th>
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<tr>
<td>R</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>L</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
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<table>
<thead>
<tr>
<th>Pachy center (µm)</th>
<th>R</th>
<th>L</th>
</tr>
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<tbody>
<tr>
<td>Mean±SD</td>
<td></td>
<td></td>
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<tr>
<td>Pachy apex (µm)</td>
<td>R</td>
<td>L</td>
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<tr>
<td>Mean±SD</td>
<td></td>
<td></td>
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</table>

<table>
<thead>
<tr>
<th>Pachy thinnest (µm)</th>
<th>R</th>
<th>L</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean±SD</td>
<td></td>
<td></td>
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</tbody>
</table>

K<sub>m</sub>: Maximal keratometry; CV: Corneal volume; ACV: Anterior chamber volume; ACD: Anterior chamber depth; Jo-1: Histidyl-tRNA synthetase antibody; ANF: Anti-nuclear factor; β2GPI: Anti-β2 glycoprotein.

### Table 5: Association between anterior segment parameters and extramuscular features of IIM patients

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Dysphagia</th>
<th>Arthritis/arthralgia</th>
<th>Weight loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>R</td>
<td>+</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>L</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

K<sub>m</sub>: Maximal keratometry; CV: Corneal volume; ACV: Anterior chamber volume; ACD: Anterior chamber depth.
values, however, there was no significant difference for OSDI scores between patients and controls. Investigating IIM patients together only few significant differences were noticed between distinct anterior segment quantities and antibodies. However, numerous significant differences were discovered between anterior segment parameters and extramuscular manifestations of IIM. A close connection was revealed between pachymetric data for both sides and arthritis/arthralgia and weight loss, moreover, sporadic associations were revealed in case of other arguments. Assaying organ specific autoantibodies in autoimmune disorders can have both diagnostic and prognostic relevance. To the best of our knowledge the first publication of circulating immune complexes in IIM was by Whitaker and Engel[35]. Nowadays, laboratory testing usually shows elevated myositis-specific antibody (MSA) levels, with a diagnostic specificity exceeding 90%, and as such generally accepted to be characteristic antibodies found in PM and DM, but they can also be present in other autoimmune diseases. Anti-aminoacyl-tRNA (e.g., anti-Jo-1, anti-PL-7), anti-Mi-2 (chromodomain-helicase-DNA binding protein 4), anti-signal recognition particle (SRP), anti-transcriptional intermediary factor 1 gamma (TIF-1 γ), and anti-nuclear matrix protein 2 (NXP-2) antibodies are taken into account in this group. On the contrary anti-Sjögren’s-syndrome-related antigen A (anti-Ro60/SS-A), anti-polymyositis/sclerodermia (anti-PM/Scl), anti-Ku, and anti-U1 ribonucleoprotein (U1RNP) antibodies are reckoned among the myositis-associated antibodies (MAAs) that are not disease-specific, but related to other connective tissue disorders, mostly myositis overlap syndromes[36]. These antibodies have not been used as IIM classification criteria yet, thus maintaining a clinical-serologic gap[37].

Ocular findings in IIM are expected to be the manifestations of vascular anomalies, immune-related dysregulations, and some genetic predispositions[38]. Since cornea is an avascular tissue the role of vascular irrelevance can be excluded. As cornea has an integrated ultrastructural morphology containing Langerhans cells and thymic stromal lymphopoietin in the peripheral epithelium, DCs in the peripheral anterior stroma and a wide variety of immunoregulatory factors in the epithelium as a consequence auto-reactive T-cell mediated immune processes should definitely bear on the formation of corneal lesions in IIM[39]. Various corneal findings in IIM may be due to diverse cellular and molecular mechanisms and a weak association between anterior segment parameters and autoantibodies thus clinical parameters could be attributed to the heterogeneity of the underlying disease. Since different mechanisms contribute to the etiopathogenesis of PM and DM, diverse corneal and DED manifestations may be explained by these molecular and cellular events.

Albeit most connective tissue diseases are immune-related and broadly identified as relative contraindications to laser refractive interventions, cataract surgeries or keratoplasties as they may be accompanied by an increased rate of intra- and postoperative complications and wound healing problems, these procedures can be accomplished when there is no active ocular interference with the inherent disease or its treatment i.e. the underlying disease is well-controlled or inactive.

The group of IIM is considered a multisystem disease that affects several organs, and by default different specialists are involved. Our data could be essential for both ophthalmologists, who deal with ophthalmic conditions of autoimmune patients, and for rheumatologists and immunologists, who treat IIM patients most likely suffering from ocular lesions as well, and accordingly, ocular examinations should be included in their routine. IIM is regarded as a rare condition, however, varied incidence and/or prevalence rates may put a considerable emphasis on this kind of autoimmune disease.

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