

Summary

Systemic sclerosis (*SSc*) is an autoimmune disease characterized by microvascular abnormalities, a multifactorial complex inflammatory reaction, and widespread fibrosis affecting the skin and various organs. The ocular adnexa and the anterior and posterior segments of the eye, including the choroid, can be affected by pathological alterations during the progression of *SSc*. The choroid is comprised of a tightly packed network of blood vessels and a stroma made up of connective tissue. The advent of optical coherence tomography (*OCT*) technology and the implementation of enhanced depth imaging (*EDI-OCT*) have led to a surge in research focused on the choroid. The results of these studies have greatly contributed to our understanding of the underlying mechanisms behind alterations in choroidal morphology associated with various retinal, choroidal, and systemic diseases.

This study aimed to investigate the pathogenesis of choroidal lesions and to assess their relationship with clinical parameters in patients with *SSc*. Understanding this relationship could help in evaluating the activity and stage of the disease. The analysis focused on the macular and peripapillary areas of the choroid. For the macular area, the thickness and volume of the choroid were measured within a 6 mm diameter, in each subfield of the conventional Early Treatment of Diabetic Retinopathy Study grid (*ETDRS*). For the peripapillary area, measurements of the retinal nerve fiber layer (*RNFL*) and peripapillary choroidal thickness were included in the analysis. Furthermore, the study incorporated a novel quantitative parameter for assessing both components of the choroid: the choroidal vascularity index (*CVI*). This comprehensive evaluation of choroidal parameters fills a gap in the literature and advances our understanding of the underlying mechanisms of choroidal involvement in *SSc*.

This was a prospective single-center, cross-sectional study conducted in the Ophthalmology Department of the Medical University of Białystok. The study involved 33 *SSc* patients (66 eyes) admitted to the Department of Rheumatology and Internal Diseases of the Medical University of Białystok. Based on the 2013 ACR/EULAR (*American College of Rheumatology / European League Against Rheumatism*) *SSc* criteria, patients were diagnosed with diffuse *SSc* (*dSSc*) or limited *SSc* (*lSSc*). The control group was composed of 40 patients (80 eyes). The groups did not differ with regard to age, sex, and axial length. All participants underwent ophthalmological

examination and OCT (*SD-OCT, Heidelberg Engineering GmbH, Heidelberg, Germany*) of the retina and choroid was performed. Data regarding age, sex, disease duration, smoking status, and medication were recorded. History of digital ulcers, the presence of interstitial lung disease, cardiac involvement, and joint involvement were also included in the analysis. Nailfold capillaroscopy was performed, and patients were stratified according to capillaroscopic pattern features as early, active, or late SSc, as proposed by Cutolo et al.

Significantly lower choroidal thicknesses were found in the macula of patients with SSc. Subfoveal choroidal thicknesses (*SFCTs*) were also significantly lower than those of the control group ($p < 0.05$). Consequently, significantly lower choroidal volume values were observed ($p < 0.05$). CVI value was significantly higher in patients with SSc, while total choroidal area (*TCA*), luminal area (*LA*), and stromal area (*SA*) values were lower in the study group than in the control group ($p < 0.05$). It is worth noting that differences in SA value were particularly pronounced. There were no significant differences in choroidal parameters between the dSSc and ISSc groups. These parameters also did not differ in the eyes of patients stratified according to early, active, or late capillaroscopic patterns. While a significantly lower pCVI (*peripapillary CVI*) value was found in patients with SSc ($p < 0.001$), no significant differences were observed in peripapillary choroidal thickness and RNFL thickness between the study groups. No significant differences in pCVI were found between the dSSc and ISSc groups ($p > 0.05$). In addition, no clinically significant relationship was found between changes in the choroidal morphology in the macula and peripapillary region and clinical parameters that could reflect disease stage and activity in patients with SSc.

To summarize, the observed alterations in the morphological parameters of the choroid confirmed choroidal impairment in patient with SSc. A higher CVI value in the macular area may indicate that stromal involvement predominates over the vascular component. This finding provides novel insights into the thinning of the choroid in patients with SSc. In contrast, a statistically significant decrease in peripapillary CVI is likely due to a decrease in the vascular layer, which could partially explain the increased susceptibility to glaucoma, particularly normal-tension glaucoma, in SSc patients.