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Macular microangiopathy in sickle cell disease using optical coherence tomography angiography

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Purpose:

To characterize the optical coherence tomography angiography (OCTA) appearance of the perifoveal macular microvasculature in visually asymptomatic patients with sickle cell disease, and to compare these findings with those of fluorescein angiography (FA).

Methods:

This is a retrospective observational case series. Eighteen eyes of 9 consecutive patients with a median age of 41 years (range: 19-54) with electrophoretic confirmation of sickle cell disease were included and analyzed. A complete ophthalmologic examination was performed, including fundus examination, FA (Spectralis HRA+OCT, Heidelberg Engineering, Heidelberg, Germany), and OCTA (RTVue XR Avanti, Optovue Inc, Fremont, California, USA). Nine eyes of five healthy subjects were also analyzed with OCTA to serve as a control group.

Results:

OCTA demonstrated microvascular abnormalities in the perifoveal region of the macula in all eyes, whereas FA appeared normal in 9/18 eyes (50%). Most capillary abnormalities were located in the temporal juxtafoveal region and involved both the superficial and the deep capillary plexuses.

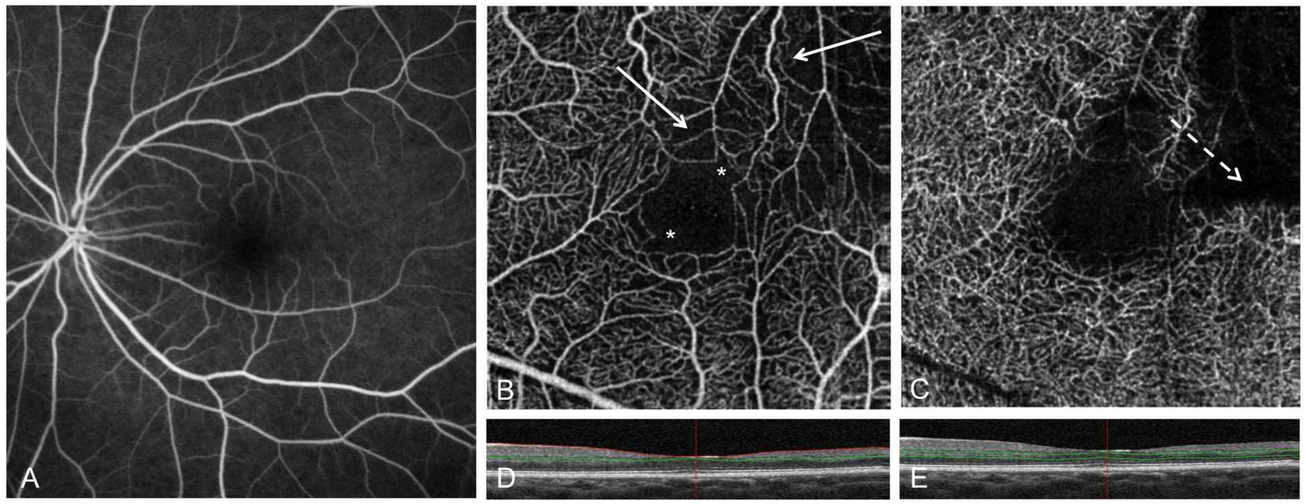

FIGURE 3. Macular microvascular changes on optical coherence tomography angiography in the left eye of a 35-year-old patient with sickle cell disease, without changes on fluorescein angiography. (Left) The fluorescein angiography frame centered on the macula shows a normal aspect of the macular area. (Top middle and Top right) The optical coherence tomography angiography frames show microvascular abnormalities in the perifoveal region at the level of the superficial (Top middle) and of the deep (Top right) capillary plexus: large areas of rarefied and dilated capillary (white arrows); areas of capillary nonperfusion (dotted arrow), maximum in the superior temporal region of the perifoveal area; disruption of the perifoveal anastomotic capillary arcade (asterisks) (best seen in the superficial capillary plexus); and enlargement of the foveal avascular zone. (Bottom middle and Bottom right) The optical coherence tomography B-scans show the automated segmentation of the superficial (Bottom middle) and the deep (Bottom right) capillary plexuses.

TABLE 1. Median Nonflow Area and Parafoveal Vessel Density in the Superficial and Deep Capillary Plexuses in the Sickle Cell Disease Group and in the Control Group

	SCD Group	Control Group	*P*
Nonflow area in the superficial capillary plexus (mm^2)	0.482 (0.346–0.827)	0.246 (0.154–0.318)	<.0001
Nonflow area in the deep capillary plexus (mm^2)	0.54 (0.334–1.214)	0.251 (0.209–0.374)	<.0001
Parafoveal vessel density in the superficial capillary plexus (%)	41.5 (30–57)	58 (42–66)	.0011
Parafoveal vessel density in the deep capillary plexus (%)	36.5 (19–58)	54 (36–69)	.0018

SCD = sickle cell disease.
Results are mean (range).

TABLE 2. Median Nonflow Area and Parafoveal Vessel Density in the Superficial and Deep Capillary Plexuses in Sickle Cell Disease Eyes With and Without Signs of Maculopathy on Fluorescein Angiography

	Maculopathy on FA (n = 9/18)	No Maculopathy on FA (n = 9/18)	*P*
Nonflow area in the superficial capillary plexus (mm^2)	0.573 (0.482–0.827)	0.419 (0.346–0.548)	.0056
Nonflow area in the deep capillary plexus (mm^2)	0.569 (0.54–1.214)	0.505 (0.334–0.574)	.0157
Parafoveal vessel density in the superficial capillary plexus (%)	37 (30–57)	42 (35–53)	.3309
Parafoveal vessel density in the deep capillary plexus (%)	29 (19–58)	38 (21–51)	.4011

FA = fluorescein angiography.
Results are mean (range).

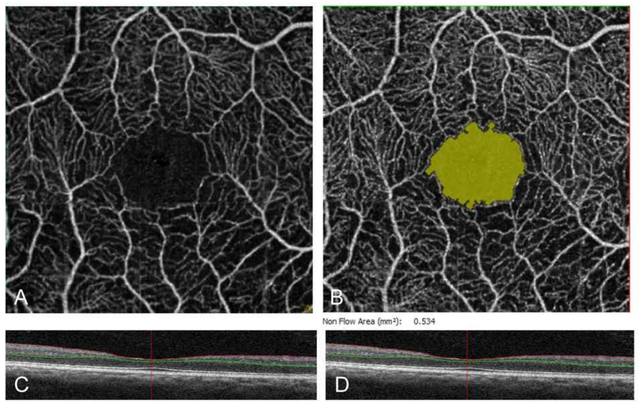

FIGURE 5. Enlargement of the nonflow area in the right eye of a 51-year-old patient with sickle cell disease. (Top) The optical coherence tomography angiography frames at the level of the superficial capillary plexus with (Top left) and without (Top right) automated delineation of the foveal avascular zone show an enlargement of the nonflow area (0.534 mm2). (Bottom) The optical coherence tomography B-scans show the automated segmentation of the superficial capillary plexus.

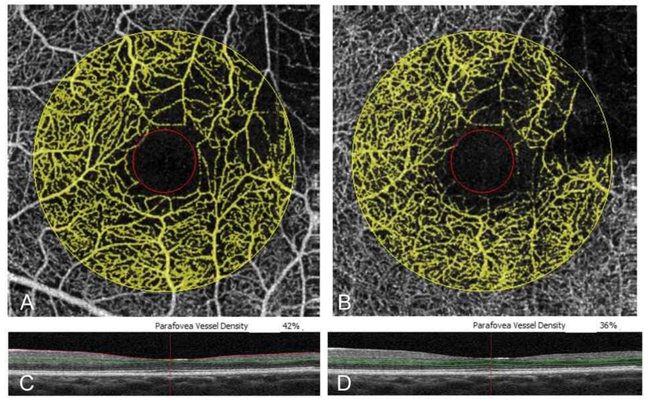

FIGURE 6. Decrease in perifoveal vessel density in the left eye of the same patient as in Figure 3. (Top) The optical coherence tomography angiography frames show a low perifoveal vessel density in both the superficial (Top left) and the deep (Top right) capillary plexuses (42% and 36%, respectively). (Bottom) The optical coherence tomography B-scans show the automated segmentation of the superficial (Bottom left) and the deep (Bottom right) capillary plexuses.

Conclusion :

OCTA provided detailed imaging of the perifoveal microvasculature in sickle cell disease. It appeared more sensitive than FA in detecting macular microangiopathy in asymptomatic patients. Microvascular abnormalities in sickle cell disease involved both the superficial and the deep capillary plexuses.