A 42-year-old man with multisystem smooth muscle dysfunction syndrome associated with a heterozygous ACTA2 gene mutation[c.536G>A, p.(Arg179His)] presented for routine ophthalmic examination. He had a history of prune belly syndrome, patent ductusarteriosus, aortic dissection, and abdominal aortic aneurysm. Best-corrected visual acuity was 20/20 in each eye. Anterior segment examination revealed mydriasis with scalloped pupillary margins, and persistent pupillary membranes extending from iris collarettes (A).Fundus examination and near-infrared imaging revealed significant retinal arteriolar corkscrew tortuosity both in the posterior pole andperiphery (B). OCT (C) and OCT angiography (D) demonstrated corkscrew vessel elongation into the outer nuclear layer.