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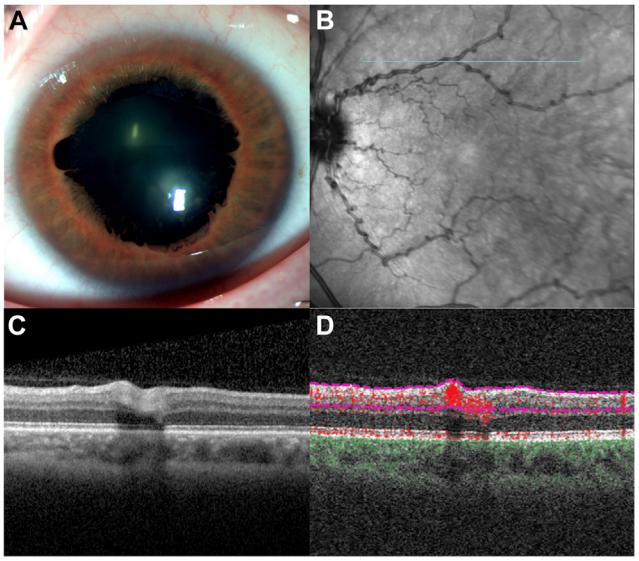
Pictures & Perspectives



Iris and Retinal Findings in Multisystemic Smooth Muscle Dysfunction Syndrome

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 ductus arteriosus, 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 and periphery (**B**). OCT (**C**) and OCT angiography (**D**) demonstrated corkscrew vessel elongation into the outer nuclear layer. (Magnified version of Figure **A**-**D** is available online at www.aaojournal.org).

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