Spontaneous Resolution of Central Serous Chorioretinopathy in a Patient With Congenital Retinal Macrovessel

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A 46-year-old man complained of sudden visual loss and a black spot in his left eye. He referred a recent stressful episode and was not using corticosteroids. Best-corrected visual acuity was 20/20 and 20/40 in the right eye and left eye, respectively. Application tonometry and anterior segment examination of both eyes were unremarkable.

Fundus examination of the left eye revealed a serous elevation of the retina in the macular area, with loss of foveal reflex. There was also a peculiar, large aberrant superotemporal venule branching inferiorly and crossing the horizontal raphe with several tributaries, two of which passed through the macula, but not involving the fovea. The right fundus was normal. Fundus fluorescein angiography (FFA) of the left eye showed a few microaneurysms in the capillary network of the perifoveal area and an ink blot pattern of leak in the temporal juxtafoveal area with diffuse spreading of the stain accumulating under the retina at the late stage with the typical aspect of central serous chorioretinopathy (CSCR) (Figure, part A).

Optical coherence tomography (OCT) revealed a serous retinal detachment (RD) of the macular area without vitreous traction (Figure, part B). The patient showed elevated 24-hour urine cortisol, although 24-hour urine catecholamines were normal. A diagnosis of acute CSCR with congenital retinal macrovessel was made. In the present case, the decreased visual acuity was secondary to the serous RD that occurred suddenly at 46 years of age, despite the congenital nature of the arteriovenous abnormality. One month later, the visual acuity in the patient’s left eye improved to 20/20. At that time, no dye leakage was detectable on FFA (Figure, part C), and the serous RD resolved spontaneously (Figure, part D).

Although the first description of an isolated enlarged vessel traversing through the macula was made by Mauthner in 1869, the term “congenital retinal macrovessel” (CRM) was coined in 1982 by Brown et al. CRM is a rare and typically unilateral abnormality of the retinal circulation, generally a single venule, which, in an aberrant manner, crosses the posterior pole or the fovea itself. It was classified in group 1 of congenital arteriovenous communications in the retina.

Prevalence of CRMs is approximately 1/200 000. Although...

Figure. A, Fundus fluorescein angiography (FFA) of the left eye showing a large aberrant superotemporal venule crossing the macula (large arrow), a few microaneurysms in the capillary network of the perifoveal area (small arrows), and an ink blot pattern of leak in the temporal juxtafoveal area (arrowhead). B, Optical coherence tomography (OCT) demonstrated a serous macular detachment due to intraretinal fluid accumulation (arrowheads). C, One month later, the visual acuity improved to 20/20, and no dye leakage was detectable on FFA. D, OCT revealed spontaneous resolution of intraretinal fluid with restoration of the foveal depression (arrowhead).
the underlying cause is unknown, it is believed these anomalies occur around the 15th to 16th week of gestation, during the differentiation and canalization of cords of mesenchymal cells invading the retinal nerve fiber layer. They are typically benign and are often detected in a routine exploration. Visual disturbance in the involved eye is infrequent and has been attributed to hemorrhage, to foveal cyst, or to the mere presence of the aberrant vessel in the foveal area.

In the present case, the decreased visual acuity was secondary to the serous RD that occurred suddenly at 46 years of age, even though the arteriovenous abnormality seemed to be congenital. A differential diagnosis must be made with other vascular entities such as vascular loops, congenital venous tortuosity or secondary to venous obstruction, arteriovenous communications, and even with tumors such as branch-shaped angioma, retinal capillary hemangioma, retinoblastoma, and choroidal melanoma. FFA proved to be valuable in establishing a definitive diagnosis. The clinical findings of unilateral neurosensory RD in the macular region and a leakage point of fluorescein that appeared early and spread over time diffusely, under the macula, are characteristic of CSCR.

Development of serous RDs and decreased visual acuity have been associated with CRM after rapid changes in gravitational forces (eg, bungee cord jumping and roller coaster rides). This combined presentation of serous RD with CRM was also attributed to vitreous traction in a case that resolved spontaneously following posterior hyaloids detachment. Recently, Kumar et al described the only case of CSCR with CRM; however, that patient was lost to follow-up. Ours is the first well-documented case report of a spontaneously resolved CSCR associated with CRM. There was no trauma, vitreous traction, or arteriovenous communication that could account for the occurrence of CSCR.

The etiologic relationship between CSCR and CRM remains to be determined. Although they could be coincidental findings, microalterations of the blood-retinal barrier might take place around the aberrant macrovessel and be associated with the anomalies of the perifoveal capillary bed and the serum levels of stress hormones such as cortisol and catecholamines typically increased in patients with CSCR.

Disclosures
None.

References
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