A 3-year-old girl presented with reduced visual acuity in the right eye with poor fixation. Her left eye was normal. No abnormal findings were observed in the anterior segment on slitlamp examination of the right eye. A relative afferent pupillary defect was present in the right eye. Fundus examination revealed a 360° radially wrinkled retina around an enlarged optic disc covered by a tuft of glial tissue. There was a central, elevated tuft of glial tissue over the optic nerve. Approximately 20 retinal vessels of differing calibers radiated from underneath the border of the glial tissue into the peripapillary retina (Figure, A). Optical coherence tomography showed detachment and proliferative change in the nasal retina (Figure, B). No systemic anomaly was found after comprehensive examination. Magnetic resonance imaging and angiography of the brain were performed to exclude basal encephalocele, dysmorphic facial features, cerebrovascular anomalies, and the PHACE (posterior fossa anomalies, hemangioma, arterial anomalies, cardiac anomalies, and eye anomalies) syndrome.