An 8-year-old female presented with a 5-month history of left visual loss. Left visual acuity (VA) (logMAR) was 20/600 (1.46), and intraocular pressure (IOP) was 16 mm Hg with timolol with an inferior bullous ERD. Optical coherence tomography showed subretinal fluid (SRF) with acoustically dense choroidal thickening on B-scan ultrasound. Indocyanine green angiography demonstrated diffuse hyperfluorescence and magnetic resonance imaging excluded leptomeningeal angiomatosis. Due to a supply interruption of verteporfin in 2021, PDT was not performed initially. Oral sirolimus was commenced to treat the ERD and port-wine stain at 2 mg daily except twice a week when she took 2 mg twice a day to achieve a stable therapeutic level of 8.0 μg/L at 7 hours postdosing. Adverse effects (headaches and mouth ulcers) were monitored on systemic review and 3-monthly blood tests (complete blood cell counts, kidney and liver function, and lipid profile) were performed.

The inferior bullous ERD resolved after 4 months. Despite complete resolution at 6 months, VA declined to 20/900 (1.66). Sirolimus was ceased at this point and 8 months later, VA improved to 20/160 (0.90). Ten months after cessation of sirolimus, SRF returned and VA reduced to 20/500 (1.40). Sirolimus was subsequently restarted at the same initial dose of 2 mg/d except 2 days of 4 mg/d per week. SRF improved by 6 weeks with complete resolution by 3 months. VA remained stable at 20/250 (1.00) at 15 months and the patient continues to receive sirolimus. B-scan measurements showed stable tumor thickness of 4.5 mm despite resolution of SRF . Timolol was ceased and IOP remained at 14 mm Hg. There was no adverse event from immunosuppression or drug intolerance during follow-up.