Failure of Systemic Propranolol Therapy for Choroidal Hemangioma of Sturge-Weber Syndrome: A Report of 2 Cases

Propranolol was incidentally discovered to induce accelerated involution of infantile cutaneous hemangioma.1 Thereafter, reports have demonstrated a favorable response to propranolol as a first-line treatment for cutaneous, orbital, and ocular hemangioma in different ages.2,3 We report the outcome of oral propranolol therapy in 2 cases with choroidal hemangioma of Sturge-Weber syndrome (SWS).

Report of Cases. Case 1. A 14-year-old girl was assessed for retinal detachment in the left eye, where vision had been poor since infancy. She had a cutaneous nevus flammeus on the left side of her face (Figure 1A). Visual acuity was 20/20 OD and hand motions OS. Intraocular pressures were 15 mm Hg OD and 32 mm Hg OS. The right eye was normal. The left showed anomalous episcleral and conjunctival vessels, dilated trabecular meshwork vessels, and total bullous retinal detachment (Figure 1B). Ultrasonography revealed a diffuse choroidal mass, of medium to high internal reflectivity and thickness of 6.3 mm (Figure 1C). The choroidal mass appeared hyperintense to the vitreous on T1-weighted magnetic resonance imaging and isointense on T2-weighted magnetic resonance imaging (Figure 1D).
imaging. These findings suggested diffuse choroidal hemangioma of SWS, producing exudative retinal detachment. No other neurological magnetic resonance imaging findings were detected. The patient and her family consented to start oral propranolol therapy to treat her choroidal hemangioma as an alternative to radiotherapy to avoid its long-term sequelae. Under cardiological supervision, oral propranolol therapy of 2 mg/kg/d was administered. The choroidal hemangioma was ultrasonographically measured every 2 months. After 6 months of propranolol therapy, there was not any discernible change in the facial angioma appearance or choroidal hemangioma size (Figure 1D) and no clinically noticeable reduction in the retinal detachment. Therefore, propranolol therapy was gradually discontinued over a month. The left intraocular pressure is controlled with topical antiglaucoma therapy. We periodically follow up the choroidal hemangioma.

Case 2. A 22-year-old man with SWS manifesting with bilateral facial nevus flammeus, leptomeningeal angiomas, and mental retardation was ophthalmologically evaluated (Figure 2A). Visual acuity was no light perception OD and 20/300 OS. He had bilateral pseudophakia and bilateral glaucoma drainage tubes in the anterior chambers (Figure 2B). The right eye appeared atrophic. The left eye revealed extensive postequatorial choroidal hemangioma without retinal detachment. Ultrasonographically, the choroidal hemangioma appeared as a diffuse mass, of medium to high internal reflectivity with 2.9 mm thickness (Figure 2C). The patient and his parents consented to oral propranolol therapy to prevent occurrence of exudative retinal detachment from the choroidal hemangioma in his seeing eye. Under cardiological supervision, oral propranolol therapy of 2 mg/kg/d was prescribed. The choroidal hemangioma was measured every 2 months with ultrasonography. Similarly, after 6 months of oral propranolol therapy, no changes were noticed in either the facial angioma color or the choroidal hemangioma thickness (Figure 2D). Subsequently, propranolol therapy was gradually discontinued.

Comment. The suggested mechanisms by which propranolol affects
hemangioma included an early effect through vasoconstriction from reduced nitric oxide release; an intermediate effect by downregulation of vascular endothelial growth factor, basic fibroblast growth factor, and matrix metalloproteinases; and a late effect through apoptosis of proliferating capillary endothelium. Guided by literature, the dose of propranolol we prescribed failed to produce any noticeable change in the ocular or cutaneous hemangioma even after 6 months of continuous treatment. Since the majority of published reports showed that the therapeutic response becomes evident a few days to weeks from initiation of therapy, propranolol was deemed ineffective in our patients. In the only report on oral propranolol treatment in SWS, an adult patient showed complete resolution of an associated exudative retinal detachment after a few weeks of therapy, contrary to the persistence of full retinal detachment observed in our case 1; nevertheless, that report did not comment on dimensional changes of the hemangioma with propranolol. Histopathologically, choroidal hemangioma of SWS has cavernous and capillary components, which might contribute to the observed different response to propranolol herein from that in capillary hemangioma. Further study is necessary to determine whether propranolol has a role in the treatment of hemangioma in patients with SWS.

Hatem Krema, MD, FRCS
Yacoub A. Yousef, MD
Priya Durairaj, MD
Ronaldo Santiago, MD

Published Online: March 28, 2013. doi:10.1001/jamaophthalmol.2013.2879

Author Affiliations: Department of Ocular Oncology, Princess Margaret Hospital/University Health Network, University of Toronto, Toronto, Ontario, Canada.

Correspondence: Dr Krema, Department of Ocular Oncology, Service 18-739, Princess Margaret Hospital, 610 University Ave, Toronto, ON M5G 2M9, Canada (htmkrm19@yahoo.com).

Author Contributions: Dr Krema had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Conflict of Interest Disclosures: None reported.