

# Denouement and Discussion

## Periocular Hemangioma: An Eye-Opening Experience

**C**linical presentation and results of ophthalmologic examination, ultrasonography, and magnetic resonance imaging were consistent with the diagnosis of a deep infantile hemangioma involving the right orbit and upper and lower eyelids, causing proptosis, lateral displacement of the globe, and complete obstruction of the visual axis. Treatment with oral corticosteroids (prednisolone; initial dose, 2 mg/kg/d) had been started at 10 weeks of age and was then increased 2 weeks later (to 2 × 6 mg/kg/d) because of inadequate response. However, the hemangioma continued growing, and by 14 weeks of age, her electroretinogram showed signs of right postretinal dysfunction. In view of this sight-threatening progress, a multidisciplinary decision was made for surgical intervention.

At the age of 17 weeks, the major bulk of the hemangioma (4.5 × 3 × 2 cm) was excised in a joint procedure by a pediatric plastic surgeon (D.D.) and an ophthalmic surgeon (Y.A.) (Figure 3). There were no postoperative complications and the baby was able to open both eyes immediately after surgery. Histopathological examination, including positive erythrocyte-type glucose transporter protein (GLUT-1) staining of the endothelium, confirmed the diagnosis of infantile hemangioma.

To prevent regrowth of the residual hemangioma, prednisolone was continued on a tapering regimen and its use was finally discontinued by the age of 7 months. Patching of the unaffected left eye was performed for 30 minutes a day. A photograph 2 months postsurgery (Figure 4) shows the excellent functional and cosmetic result. The proptosis and lateral displacement of the globe had fully resolved and, importantly, both electroretinogram and visual evoked potentials proved to be normal for each eye on repeated testing. When last seen at the age of 9 months, the patient was well and there was no rebound growth of the residual hemangioma.

Infantile hemangioma is the most common benign tumor of infancy, occurring in 4% to 10% of children.<sup>1</sup> Hemangiomas are usually absent at birth or present only as precursor lesions and have a characteristic natural history of rapid growth within the first 3 to 6 months of life, followed by spontaneous involution over several years.<sup>2</sup> Most hemangiomas are asymptomatic and can be managed by observation; however, a few may cause significant complications. Periocular hemangiomas can lead to severe and permanent visual disturbances by occluding the visual axis, compressing the globe, or expanding into the retrobulbar space. The diagnosis is usually established clinically, but imaging studies should be considered if there is

a concern regarding other orbital soft tissue tumors (rhabdomyosarcoma, neuroblastoma, plexiform neurofibroma, or lymphatic malformation) and to assess the full extent of the tumor.<sup>3</sup> During the growth phase of periocular hemangiomas, patients warrant close evaluation and follow-up by a multidisciplinary team comprising an ophthalmologist, dermatologist/pediatrician, and ideally a plastic surgeon. Systemic corticosteroids are the treatment of choice for hemangiomas that cause visual compromise. Other treatment options include intralesional or topical corticosteroids, interferon- $\alpha$ , and surgery, but their use may be limited by potential risks or lack of efficacy.<sup>3</sup> The vascular nature of these lesions poses the challenge of potentially significant hemorrhage and surgical intervention is therefore often avoided. However, as we report herein in a child with a sight-threatening periocular hemangioma who failed to respond to oral corticosteroids, surgical debulking can be a valuable treatment option to save vision with an excellent cosmetic result.

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