

## Grand Rounds

### A 7-week-old infant with right upper eyelid mass

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#### History

A 7-week-old girl presented with a right upper eyelid mass. She was born at a gestational age of 30 weeks by Cesarean section with a birth weight of 935 g. She had since been in the neonatal intensive care unit, where she was treated for pulmonary insufficiency, anemia of prematurity, and meconium plugging. The right upper eyelid lesion was deep red in color, had been present for 3 days, and was growing. Notably, on retinopathy of prematurity (ROP) rounds one week before, no right upper eyelid lesion had been appreciated.

#### Examination

On examination, the patient blinked to light in each eye independently. There was no afferent pupillary defect. Intraocular pressures were soft to palpation in each eye, and motility was within normal limits by doll's head maneuver. Visual fields could not be assessed due to the patient's age. The external examination findings are shown in Figure 1. On penlight examination, the sclerae and conjunctivae were white and quiet, the corneas were clear, the anterior chambers were formed, and the irides were round with no lesions. The lenses also appeared clear in both eyes. The dilated fundusoscopic examination revealed pink, optic nerves with sharp borders and a cup-to-disc ratio of 0.2. The maculae in each eye were flat, and the vessels were of normal course and caliber. Finally, the periphery was normal in both eyes also.

#### Ancillary Testing

The eyelid lesion did not transilluminate.

#### Treatment

The patient's family was presented with all treatment options and made aware of the concern for occlusion



Figure 1. Appearance of lesion at presentation.

amblyopia given the rapid progression of the mass. Propranolol was not advised by the primary ICU team due to concerns regarding a heart murmur and the patient's overall cardiac status. After a full discussion of the indications, risks, benefits, and alternatives, surgical excision of the right upper eyelid mass was performed (Figure 2A–D). Pathology slides are shown in Figure 3A–D.

Twelve days postoperatively, the patient was noted to have a recurrence, which continued to progress over the next two weeks. At that point (1 month postoperatively), the patient was given an intralesional corticosteroid injection (50/50 mix of 40 mg/mL triamcinolone and 6 mg/mL betamethasone). There was no recurrence at 2 months' follow-up (Figure 4).

#### Differential Diagnosis

The differential diagnosis in this case should include all of the following: hematoma, capillary hemangioma, lymphangioma, arteriovenous malformation, port-wine stain, and neuroblastoma. A hematoma may have formed due to the previous week's ROP examination.

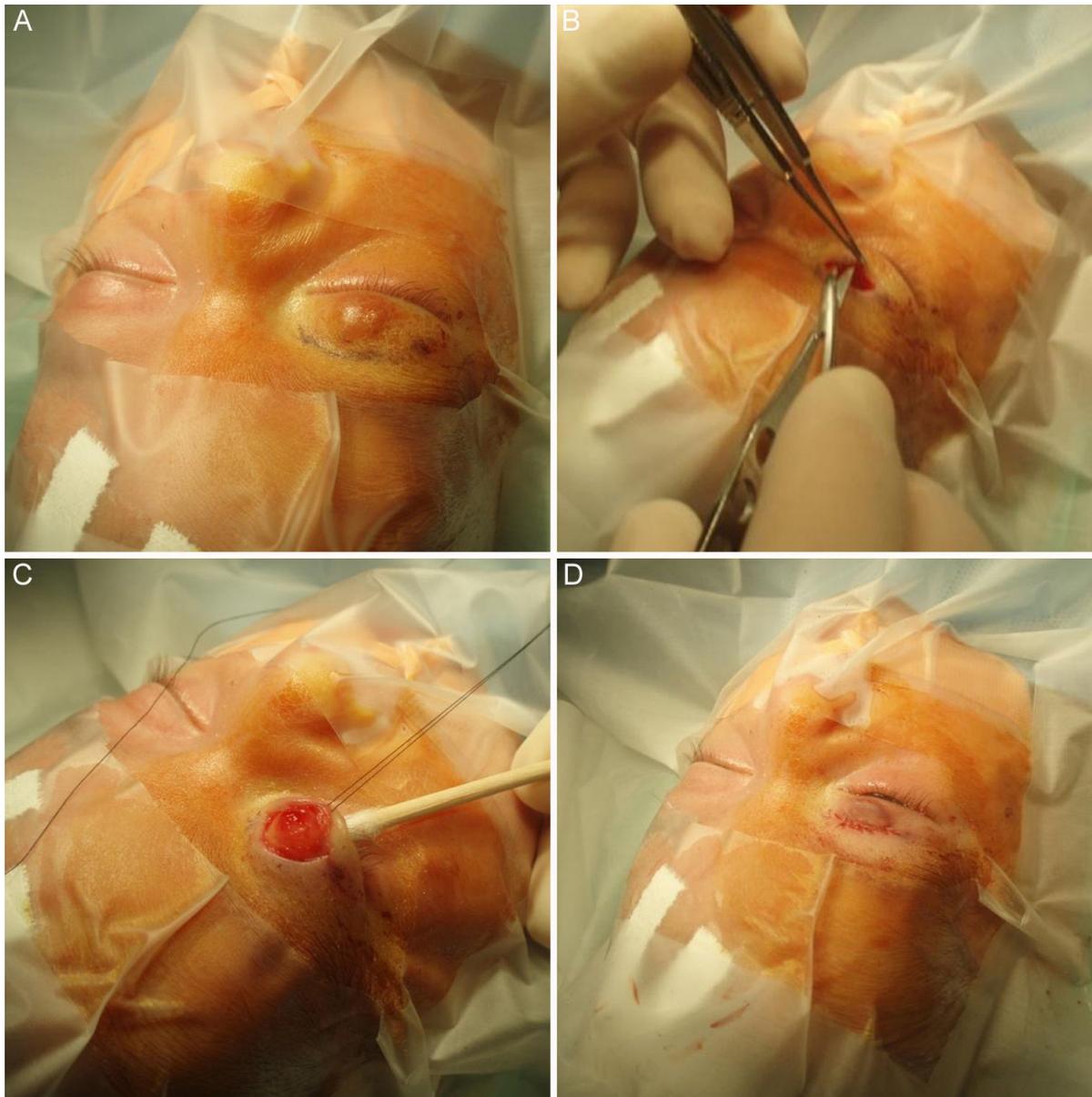
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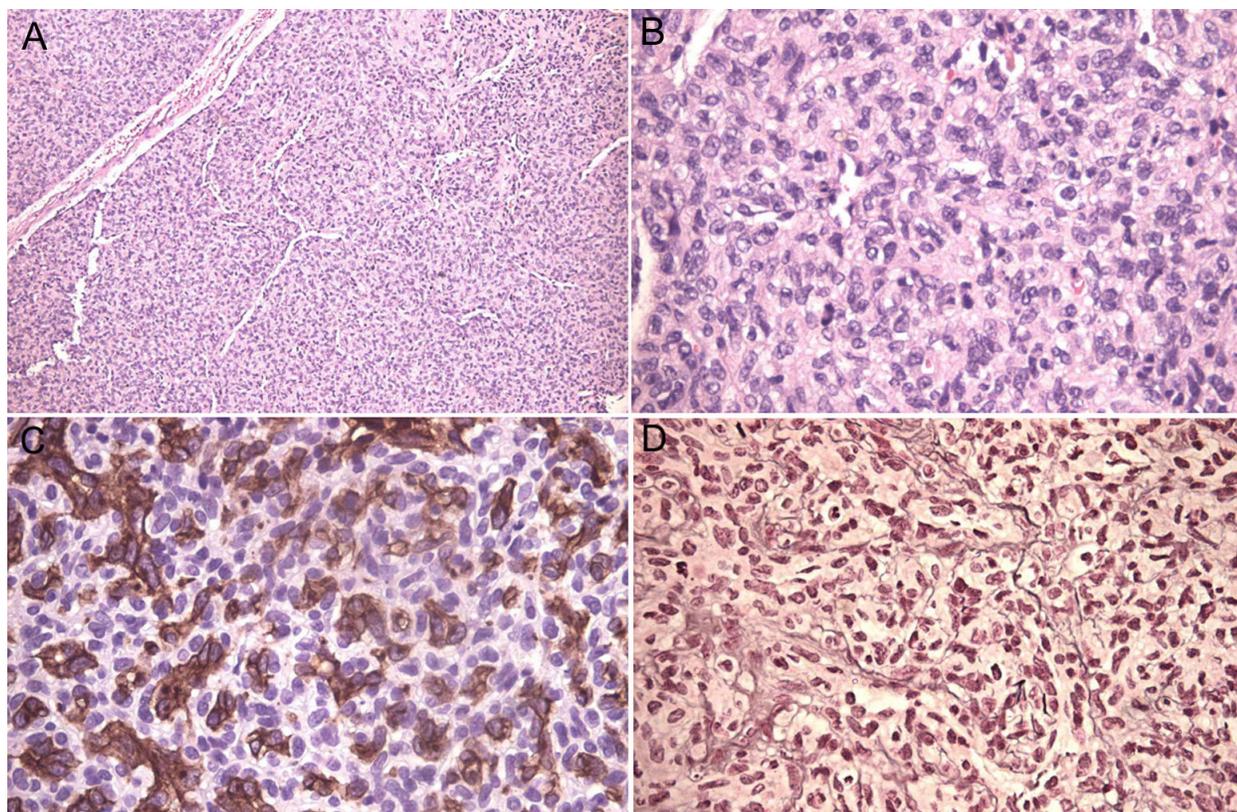
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**Figure 2.** Surgical excision of right upper eyelid lesion. A, Preoperative patient preparation, with incision marked above eyelid crease. B, Incision made along previously noted mark. C, Exposure of eyelid mass. D, Postoperative photograph after mass excision and closure.

Lid speculum placement can be traumatic in newborn babies, and a good assistant is essential to prevent unnecessary movement. Capillary hemangiomas are vascular lesions that may present shortly after birth with progressive enlargement and are more common in females and with premature birth.<sup>1,2</sup> Lymphangiomas present in the first decade of life and often present abruptly as a localized progressive vascular lesion with spontaneous hemorrhage.<sup>3</sup> Arteriovenous malformations consist of anomalous anastomoses between arteries and

veins, are typically congenital, and may present with a localized hemorrhage.<sup>4</sup> Port-wine stains (nevus flammeus) consist of deep, dilated capillaries that are often present at birth, frequently affect one side of the face, and can deepen in color as a child ages.<sup>5</sup> Neuroblastoma is the most common extracranial cancer in children and metastatic neuroblastoma can present in young children with sudden and rapidly progressive eyelid ecchymosis that may be unilateral or bilateral.<sup>6</sup>



**Figure 3.** A, Histology demonstrating high cellularity with only rare vascular components (hematoxylin and eosin, original magnification  $\times 40$ ). B, On higher magnification, numerous mitotic figures were noted, reflecting a highly proliferative lesion (hematoxylin and eosin,  $\times 200$ ). C, A CD31 stain emphasizing the numerous endothelial channels in the lesion ( $\times 200$ ). D, Positive reticulin stain surrounding capillary vessels ( $\times 200$ ).

## Diagnosis

The diagnosis in this case was capillary hemangioma as evidenced by the clinical and histological appearance of the lesion. Capillary hemangiomas, previously known as strawberry hemangiomas, are hamartomatous growths of vascular endothelial cells.<sup>1</sup> They are not generally present at birth but appear within the first few weeks of life, reaching their peak size at approximately 6–12 months before decreasing in size over the next 4–5 years. Many of these capillary hemangiomas spontaneously resolve, with studies showing resolution of 50% by 5 years and 80% by 8 years. Ophthalmological indications for treatment include occlusion of visual axis or induced astigmatism, leading to concern for amblyopia.<sup>2</sup>

Numerous treatments exist for capillary hemangiomas. Small non-elevated lesions can frequently be observed. First-line treatment for larger lesions typically includes corticosteroids, which can be administered topically (0.05% clobetasol cream) for superficial lesions, by



**Figure 4.** Patient 2 months postoperatively. Note the surgical scar visible on the right upper eyelid above the lid crease. There is no evidence of recurrence.

intralesional injection (40mg/mL triamcinolone and 6mg/mL betamethasone) for intermediate or periorbital lesions, and orally (2mg/kg/day) for refractory lesions.<sup>7</sup> Corticosteroids carry significant risk, with systemic corticosteroids potentially causing growth retardation, personality changes, infections, adrenal crisis, diabetes risk, and even rebound growth on cessation.<sup>8</sup> Topical clobetasol cream minimizes many of these risks but still causes dermal atrophy and pigmentary changes, and it can take several weeks to achieve a therapeutic response.<sup>8</sup> Intralesional corticosteroid injection is often used as a first-line treatment, although it adds the risk of central retinal artery occlusion, which has been reported in the contralateral eye as well.<sup>8</sup>

Pulsed-dye laser therapy has been shown to successfully treat flat superficial lesions or residual surface blood vessels after steroid treatment.<sup>9</sup> This therapy has been particularly useful in cosmetically significant areas, including the periocular region. Surgical excision may be employed in refractory cases, particularly with lesions that appear well circumscribed. These lesions are not typically encapsulated, so there is a potential for recurrence as well as a risk of bleeding and a risk associated with general anesthesia.<sup>9</sup>

Interferon alpha-2a, which has been shown to inhibit angiogenesis and endothelial cell migration and proliferation in vitro, has shown promise in refractory cases as well; however, its use has adverse effects, including motor developmental delay and spastic diplegia.<sup>10</sup>

Propranolol has shown great promise in the treatment of capillary hemangiomas.<sup>11,12</sup> In one study, 11 children with capillary hemangiomas were administered 2mg/kg/day of propranolol. Many of these patients began exhibiting color changes of their lesions within 24 hours, and all patients experienced resolution of their capillary hemangiomas, although varying treatment intervals were required.<sup>11</sup> It has been hypothesized that

propranolol causes vasoconstriction of this highly vascular lesion and acts to down-regulate vascular endothelial growth factor and basic fibroblastic growth factor (bFGF), eventually triggering apoptosis of capillary endothelial cells.<sup>11</sup>

## References

1. Haik BG, Karcioğlu ZA, Gordon RA, Pechous BP. Capillary hemangioma (infantile periocular hemangioma). *Surv Ophthalmol* 1994;38(5):399-426.
2. Schwartz SR, Kodsi SR, Blei F, Ceisler E, Steele M, Furlan L. Treatment of capillary hemangiomas causing refractive and occlusion amblyopia. *J AAPOS* 11(6):577-83.
3. Harris GJ, Sakol PJ, Bonavolonta G, De Conciliis C. An analysis of thirty cases of orbital lymphangioma: pathophysiologic considerations and management recommendations. *Ophthalmology* 1990;97(12):1583-92.
4. Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions. *Ophthalmology* 2004;111:997-1008.
5. Li G, Lin T, Wu Q, Zhou Z, Gold MH. Clinical analysis of port wine stains treated by intense pulsed light. *J Cosmet Laser Ther* 2010;2(1):2-6.
6. Miller, NR., editor. *Walsh and Hoyt's Neuro-Ophthalmology*. 4th ed. 3. Baltimore: Williams & Wilkins; 1988. p. 1296-1300.
7. Ranchod TM, Frieden IJ, Frederick DR. Corticosteroid treatment of periorbital haemangioma of infancy: a review of the evidence. *Br J Ophthalmol* 2005;89(9):1134-8.
8. Boon LM, MacDonald DM, Mulliken JB. Complications of systemic corticosteroid therapy for problematic hemangioma. *Plast Reconstr Surg* 1999;104(6):1616-23.
9. Azizkhan RG. Laser surgery: new applications for pediatric skin and airway lesions. *Curr Opin Pediatr* 2003;15(3):243-7.
10. Barlow CF, Priebe CJ, Mulliken JB, et al. Spastic diplegia as a complication of interferon alfa-2a treatment of hemangiomas of infancy. *J Pediatrics* 132(3):527-30.
11. Léauté-Labrèze C, Dumas de la Roque E, Hubiche T, Thambo JB, Taïeb A. Propranolol for severe hemangiomas of infancy. *N Engl J Med* 2008;358(24):2649-51.
12. Cruz A, Siegfried EC. Propranolol treatment for periocular capillary hemangiomas. *J AAPOS* 2010;14(3):199-200.