CLINICAL VIGNETTE

Retro-Orbital Infantile Hemangioma: A Rare Cause of Proptosis

Kimberly Dang, MD, Kelsi McCoy-Wilson, MD and Indu Kannan, MD

Introduction

We present a 2.5-month-old, full term female infant presenting with an unusual orbital complaint. She was large for gestational age, with normal spontaneous vaginal delivery. Her mother's pregnancy course was complicated by gestational diabetes mellitus controlled with metformin, DVTs on prophylactic enoxaparin, and asthma controlled with budesonide. The postnatal course was complicated by hypoglycemia requiring a brief NICU stay for IV fluids, with subsequent discharge without any major issues. Her growth parameters at birth were 94 percentile for weight, 80 percentile for length and 88 percentile for head circumference. She was healthy until 2 months of age when she presented to her pediatrician with new onset, progressively worsening proptosis of the right eye without other symptoms (Figure 1). The proptosis was first noted by her mother 4 days prior to presentation. She was without excessive right eye tearing, light sensitivity, erythema, fever, or vomiting. The patient was referred to the Emergency Department and was subsequently admitted for expedited work up and ophthalmologic consultation.

Figure 1
Patient Pictures - Left image is the patient prior to treatment, note right sided proptosis with increased scleral show. Right image is after approximately 1-2 weeks of propranolol treatment with near complete resolution of proptosis.

On presentation, physical exam was notable for significant right sided proptosis but otherwise normal appearing external eye exam. Ophthalmologic evaluation demonstrated normal right eye intraocular pressure and equivalent K diameter. Ocular motility examination, revealed a supraduction deficit of -3 in the right eye. Dilated fundus exam was normal but there was concern for a subretinal temporal mass and MR imaging was recommended. MRI Brain and Orbits with and without contrast revealed features consistent with a rapidly growing retro-orbital infantile capillary hemangioma that was displacing and compressing the intraorbital optic nerve (Figure 2). Given the rapid growth, severe degree of proptosis, and concern for impending compressive optic neuropathy, the patient was started on systemic propranolol. Propranolol was initiated at 1mg/kg PO divided TID and increased to 2 mg/kg PO divided TID prior to discharge. The patient was seen in clinic 1 week after propranolol initiation with dramatic response and near resolution of proptosis. The patient tolerated propranolol well without hypoglycemia, bradycardia, or hypotension. The patient will continue propranolol for at least 6-12 months with close follow up with ophthalmology and primary care.

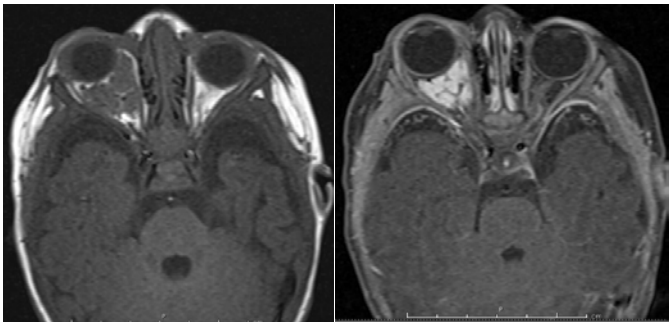

Figure 2
MRI Brain and Orbits with Contrast - Left image is T1 weighted image demonstrating right retro-orbital lesion with isointensity to muscle. Right image is T2 weighted image with hyperintense right retro-orbital lesion with vascular flow voids and avid enhancement. The retro-orbital mass is multilobulated and occupies much of the inferior intraconal space and extends extraconal and preseptal along the inferior margin.

Discussion

Differential Diagnosis of the Orbital Masses

It is of great importance to differentiate different types of orbital masses from one another in children as treatment varies significantly by causality. Orbital masses can be differentiated by location: eyelid, ocular surface, orbit, or intraocular. They can be vascular lesions, inflammatory masses, cysts, hematopoietic lymphoproliferative lesions, or neurogenic tumors.[1] The most common vascular lesions of childhood are capillary hemangio-

mas, cavernous hemangiomas, AV malformations, and lymphangiomas.[1]

Infantile Hemangiomas

Capillary hemangiomas, which are also referred to as infantile hemangiomas (IH), are the most common benign tumors of childhood, occurring in 4-5% of infants.[2] They can be superficial, intradermal, or subdermal. Superficial hemangiomas are usually bright red and bumpy in appearance, while subdermal lesions present as blue masses.[1] IH develop in the first few weeks or months of life, with most rapid phase of growth occurring in the 1st year of life. Approximately 80% of growth occurrs by 3 months of age, followed by an involuting phase lasting 1-7 years, and expected complete regression by 8-12 years.[3] IH can occur in a variety of cutaneous regions including the face, trunk, extremities, and deeper structures such as the liver, airway, and orbit.[2] Risk factors for development of IH include genetic predisposition and imbalance of angiogenic factors.[3] Complications include high output congestive heart failure, thrombocytopenia, nasopharyngeal or supraglottic obstruction, bleeding, dermal ulceration, and permanent scarring.[4]

IH of the orbit are rare and require immediate evaluation due to the potential for permanent visual impairment. These hemangiomas can cause visual axis occlusion, strabismus, ptosis, proptosis, corneal exposure, and compressive optic neuropathy.[4] There have been reports of retro-orbital IH causing congenital glaucoma.[3] Characteristics of the hemangioma may prompt evaluation for possible posterior fossa malformations, arterial anomalies, cardiac defects, eye abnormalities, sternal cleft and/or supraumbilical raphe (PHACE) syndrome.[2]

Management

Management of IH depends on the location, degree of functional impairment or ulceration, or presence of life-threatening complications.[2] About 10% of IH require treatment with orbital capillary hemangiomas, the goal of treatment is vision preservation.[3] If there is no or minimal visual impairment, close observation can be considered. There are several treatment options with variable efficacy and safety profiles. Oral propranolol is the treatment of choice and been used to effectively treat IH, both of the orbit and elsewhere on the body.[5] Its mechanism of action is via decreasing expression of growth factors and hypoxia induced capillary endothelial cell apoptosis.[3] Propranolol is generally well tolerated, but adverse reactions include transient hypoglycemia, hypotension, bradycardia, and bronchospasms.[5]

A similar case of a 4-month-old female with isolated orbital IH who was treated with propranolol and demonstrated complete resolution on repeat imaging 3 months after initiation of treatment. Propranolol was continued for a total of 8 months.[5] Our patient received propranolol with a dramatic response resulting in near resolution of proptosis 1-week post treatment initiation as seen in Figure 1.

IH may also be treated with topical, intralesional, or systemic steroids. The mechanism of action is thought to be sensitization of the hemangioma vasculature to circulating vasoconstrictors.[4] Other less commonly used treatments are carbon dioxide lasers, interferon alpha, vincristine, cyclophosphamide, embolization, and surgery.[6] However, these therapies have higher risk of adverse reactions, such as neurotoxicity with interferon alpha, injury to the optic nerve and intraocular muscle with embolization and surgery, and a multitude of systemic effects with vincristine and cyclophosphamide.[1,7] In contrast to superficial hemangiomas that can be monitored clinically, deeper hemangiomas, such as orbital hemangiomas, may require repeat imaging for monitoring.[5]

Conclusion

We present a patient with the uncommon diagnosis of retro-orbital infantile hemangioma, presenting with unilateral proptosis. She responded rapidly to a noninvasive treatment with oral propranolol, which was well tolerated without adverse effects. When encountering a pediatric patient with unilateral proptosis, it is important to have a broad differential diagnosis and perform a thorough evaluation that includes imaging and ophthalmologic consultation for vision preservation.

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