

Article

The Young Child with Capillary Hemangioma: Case Report and Review

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ABSTRACT

Background: Capillary hemangiomas are a frequently diagnosed pediatric vascular lesion. These hemangiomas are caused by a proliferation of large, aggregated endothelial cells that have few capillary lumina. They present shortly after birth and are noninheritable benign tumors. The hemangiomas usually grow rapidly for approximately one year and then undergo spontaneous involution. These benign tumors interfere with eye growth and threaten vision by occluding the eye. Many infants develop amblyopia, anisometropia and strabismus.

Case Report: A 4-month-old black female (BI) presented with a large capillary hemangioma covering her left eye and cheek. She had been taking oral prednisone since one month of age. The hemangioma appeared to completely occlude her eye, but when lifted an intact eye was noted. The patient's mother was interested in having the lesion removed at this time. A comprehensive therapy plan was instituted to help provide visual stimulation and to decrease the possibility of developing amblyopia. Unfortunately, the patient was lost to follow-up until 4 years of age. BI had had the capillary hemangioma removed at 3 years of age. BI was

diagnosed with deprivation amblyopia and esotropia. Patching therapy was initiated. Patient cooperation unfortunately was poor.

Conclusion: Since the American Optometric Association recommends that children receive their first comprehensive eye and vision examination at 6-12 months of age and since the College of Optometrists in Vision Development has always supported early intervention, the pediatric/functional/behavioral optometrist should be ready to diagnose and manage a wide range of disorders including those whose etiology may be pathological in nature. This case demonstrates the importance of the prevention of occlusion amblyopia if surgical removal of the capillary hemangioma is not indicated. BI was not a candidate for surgical removal because of her initial age. These tumors frequently resolve or decrease in size within the 1st year or so. This case also illustrates the importance of appropriate patient follow-up and cooperation and the consequences of what can occur when that cooperation is lacking.

KEY WORDS

capillary hemangioma, tumor, occlusion amblyopia, therapy

INTRODUCTION

Capillary hemangiomas are the most common benign tumors found in children. These tumors show no pattern of inheritance from either parent, nor specific ethnic or national preference.¹ Capillary hemangiomas are caused by a proliferation of large, aggregated endothelial cells and anastomosing blood filled channels. They present shortly after birth, with a predilection for females and have an affinity for the superior lid. The growth of these tumors is most active during the four months after its initial appearance but

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Figures 1 and 2. Capillary Hemangioma



Figure 3. Eye underneath

often continues over an eight month period.² Spontaneous resolution of the tumor is seen in many patients. Forty percent resolve by the time a child is four years old and 70% resolve by the age of seven.³

Capillary hemangiomas of the eye and orbit may interfere with the growth of the globe and influence its final shape. When a hemangioma threatens vision by occluding a portion of the eye or the entire eye, occlusion induced amblyopia becomes a concern. These infants are at risk for developing amblyopia, anisometropia and strabismus.⁴ This paper discusses a case in which a hemangioma caused complete occlusion of a young patient's left eye. A review of the management options is discussed as well.

CASE REPORT

A four month old black female (BI) in minimal apparent distress presented to the Illinois Eye Institute. BI's mother had taken her to a hospital concerning a lesion on the infant's face. At this visit she was referred to another hospital for further evaluation. The second hospital told her the lesion could not be removed at this young age. BI was prescribed oral prednisone twice a day at one month of age. The patient's mother wanted to have the lesion removed and came to the Illinois Eye Institute for a second opinion. (See Figures 1-3.)

The patient's ocular history revealed no trauma or surgery. Her medical history was significant for a premature birth. BI was born at 25 weeks with a birth weight of 1 lb 13 ounces and was on postnatal oxygen. Lesion growth began one month after birth and had been increasing in size since that time.

Visual acuity assessment revealed the right eye was able to fixate and follow light. The extraocular muscles in the right eye showed a full range of motion. While the left eye was able to fixate, the large lesion made it challenging to determine if she was actually following the light into the various positions of gaze. Her pupils were equal, round and reactive to light in both eyes. The Hirschberg/Kappa tests revealed an esotropia of

approximately 15 prism diopters in the eye with the hemangioma.

The mother's main goal was to have the tumor removed for cosmetic reasons. We explained that aside from cosmesis, BI was in danger of occlusion amblyopia secondary to the hemangioma. We reviewed several visual stimulation techniques and how to perform these procedures at home with the mother. An appointment was made for BI with our pediatric ophthalmologist. The ophthalmologist felt that she should continue the oral steroids and was to be followed until the tumor regressed. We scheduled appointments for the patient to have vision stimulation therapy. Unfortunately, BI did not return for follow up care at that time.

Four years after her initial evaluation, BI returned for a comprehensive examination. (See Figure 4). She had had the capillary hemangioma removed at 3 years of age. Her visual acuity (VA) was 20/25 OD using Allen symbols. The visual acuity for the left eye using Teller Acuity showed 20/63 at 55cm. Stereopsis was not present. The Hirschberg and Krimsky tests revealed a 20 PD esotropia OS. Retinoscopy showed +0.25 Sph in OD and Pl -4.00 X 090 in OS. She was diagnosed with deprivation amblyopia and patching therapy was initiated. Patient cooperation unfortunately was poor and her visual acuity 3 months later had not improved.

DISCUSSION

It is important to rule out any life or sight threatening diseases when diagnosing capillary hemangiomas. Capillary hemangiomas may or may not cause visual dysfunction. The following table (Table 1) presents the differential diagnosis.

Many secondary problems occur when a capillary hemangioma involves the eyes. The most serious problem that can occur is that vision may not develop properly and amblyopia results. For infants, even brief occlusion of the eyes (especially in the first year of life) may be harmful for visual development. Form deprivation is one of the main causes of severe



Figure 4. Capillary hemangioma removed Side view.

amblyopia in these infants. Good visual acuity can result if the tumor regresses or is removed by the third month of the infant's life.⁷ Amblyopia patching therapy can also be initiated once the eye is no longer occluded.

Deprivation amblyopia is of major concern in these patients. In order for visual acuity to develop, it is vital to address the problem immediately. Unfortunately, the hemangioma could not be removed from BI without risking her life. Deprivation amblyopia is secondary to inadequate retinal stimulation due to form distortion that often results from an obstruction of the ocular media.⁸ Monocular congenital cataracts are the leading cause of this type of amblyopia.⁹ Ptosis, corneal and vitreal opacities as well as tumors on the eyes can all cause form deprivation amblyopia.

The development of anisometropia can also lead to amblyopia. There is usually a greater amount of myopia in the affected eye.⁷ Permanent refractive errors can be caused by pressure of the tumor. The hemangioma usually exerts pressure on the eye in a direction perpendicular to the axis of the astigmatism. If the tumor regresses early or is removed, the induced astigmatism will often return to normal. Patching therapy should be implemented as soon as possible for infants at risk of amblyopia. Haik et al suggest patching the preferred eye

all but two waking hours daily when beginning treatment. Weekly follow ups should be done to monitor the patient's visual acuity. As acuity improves, patching time should be decreased slowly.

Strabismus can also occur in the affected eye. The hemangioma can affect muscle function directly by involving the muscle. As the tumor regresses, the strabismus also begins to resolve.⁷ For this reason, strabismus surgery is delayed until the tumor shrinks significantly or is removed. We suggested that instead of full time occlusion, the preferred eye be patched for fifteen minute intervals six times a day. While the eyelid was held open, BI's mother was to present visually stimulating objects to the affected eye. She was instructed to use flashlights, Christmas lights or illuminated toys. It was explained that BI's left eyelid would have to be held open while showing her the light stimulus. While showing the light it should be moved in different fields of gaze and the child should be encouraged to look at the light.

Treatment Options:

There are many treatment options available for capillary hemangiomas. The most common are observation without treatment and intralesional steroids. These are discussed in Table 2.

The observation without treatment option was not chosen for BI because the tumor was large and occluded the eye. She needed immediate treatment to decrease her chances of developing occlusion amblyopia, therefore oral steroids were utilized. Surgical excision was not chosen because of her age and the active growth of the tumor. The hemangioma was very large and surgery is typically performed on smaller lesions. She would be at risk for excessive bleeding because the tumor was deeply embedded in the epidermis. The LASER option was not used because of the tumors large size. Interferon alpha 2-a was never suggested as an option to the parents.

Table 1: Differential Diagnosis^{4,5,6}

	Age of onset	Benign/ Malignant	Proptosis	Treatment	Comment
Capillary Hemangioma	Birth to 6 months	Benign	Can be seen in some cases	Can resolve on its own. Laser tx, Surgery, steroids, Interferon 2a	Usually dx clinically. Proliferation of large endo- thelial cells
Lymphangioma	Usually presents in first decade of life	Benign	After undergoing spontaneous hemorrhage	Can be removed surgically; may reoccur	Cystic lymphatic mal-formation that occurs in orbit
Rhabdomyosarcoma	Within first decade of life	Malignant and may metastasize	Rapidly expanding globe with proptosis	Surgery, chemo tx, radiation	Must undergo biopsy. Rapidly growing
Dermoid cysts	Birth to young adulthood	Benign	No proptosis is seen	May require surgery to remove	Asymptomatic firm and round localized mass
Orbital Cellulitis	Present at any age	Can be sight threatening	Proptosis is part of the diagnosis	Oral antibiotics	Lid edema, erythema. Hx of fever or sinus infection

Observation without Treatment	40% of lesions resolve by age four; 70% resolve by age seven	Tumors reach full size by 6 months to 1 year	Cosmetic repair should be delayed until 8 years of age (when tumor becomes inactive)	Excellent chance of spontaneous regression if tumor is small.
Oral Steroids	Main reason for treating patients with steroids is to try to prevent amblyopia	A response is usually seen within two weeks	Mechanism of action is unclear, it may be secondary to a vasoconstrictive affect	Various side effects include growth delays, Cushing's disease and an increased susceptibility to infections
Intralesional Corticosteroids	Most commonly used and promising treatment for capillary hemangiomas	Best if there is direct injection into the lesion because it will have minimal systemic absorption	Multiple injections often required	Side effects include retrobulbar hemorrhage, central retinal artery occlusion and eyelid necrosis
Surgical Excision	Performed during the inactive phase, when the tumor is least likely to grow (after the age of 8)	Surgery not used for massive tumors or those that extend into the dermis because of the risk of excessive bleeding	May leave scars	Lesion may not be removed in time to prevent occlusion amblyopia
Radiation	Treatment not commonly used	Low possibility of oncogenesis	Should only be used when a risk of deformity is present and alternative treatment not available	Tumor size decreases in 1-2 weeks
LASER Therapy	Carbon dioxide, argon, neodymium YAG lasers used	Carbon dioxide laser coagulates small blood vessels while cutting through the vascular tumor	Most effective when the tumor is diagnosed	Lasers are thought of as experimental and not as a routine method of treatment
Interferon alpha 2-a	Newest development for treatment. Injectable	Reserved for life threatening or sight threatening corticosteroid resistant hemangiomas	Success rate is approximately 50% with regression occurring after 8 months of age	Response is slow and usually not fast enough to stop occlusion amblyopia

	4 Months Old	4 Years Old
Vision	OD: Fixates and Follows light OS: Fixates, but did not follow	OD: 20/25 with Allen Symbols OS: 20/63 @ 55cm with Teller Acuity
Stereo	Unable to test	No Stereopsis with Lang Test
Eye Posture	15Δ Left Esotropia	15-20Δ Left Esotropia
Rx Given	None. Recommended Amblyopia therapy	OD: +0.25 Sph OS: PL -4.00 X 090 Discussed CL option and continuation of Amblyopia therapy

CONCLUSION

Optometrists should educate parents on what capillary hemangiomas are and to offer information on support groups for parents who have children with tumors. These support groups are shown in Table 4.¹⁵

Once the parent understands what a hemangioma is, they will better comprehend why therapy is being conducted. It is essential that parents realize that if the hemangioma occludes the eye, it can cause loss of sight if not treated.

Treatment should first begin by giving the proper prescription. This may be difficult in a spectacle Rx if the hemangioma will not allow the glasses to stay on. A contact lens may be prescribed if glasses do not fit properly. Patching therapy for amblyopia can also be initiated. The patching should be for only short periods of time so as not to cause occlusion amblyopia in the normal eye. Lastly, the patient's eye should be visually

stimulated so as to decrease the chances of developing amblyopia.

A light box can be used (available through American Publishing House for the blind) to help stimulate vision.¹⁶ The tools provided help the infant learn basic visual skills, eye hand coordination, matching and discrimination. The light box produces high contrast targets and the associated faceplates are brightly colored, moveable, and help capture the infant's attention. An example of using the light box would be by placing an infant in front of the box and noting if the infant blinks, flinches or becomes more active. All of these can mean the infant is aware of the light and box. (See Figure 5 for an example of the box and objects.)

Visual stimulation techniques can also be conducted at home. Parents can use penlights, flashlights with different colored acetate sheets in front and Christmas lights or tinsel to stimulate vision. Any object that projects light, shines and/or holds the infant's attention

Table 4: Support Groups (http://www.birthmark.org/support.html)		
The Hemangioma and Vascular Birthmarks Foundation	P.O. Box 106 Latham, NY 12110-0106 (518) 782-9637	Non-profit organization. Provides up to date information on diagnosis and treatment of hemangiomas and vascular birthmarks
Lets Face It	P.O. Box 711 Concord, MA 01742-0711 (508) 371-3186	Information and support for people with facial differences and their family and friends
Hemangioma Hope	8400 Rohl Road North East, PA 16428-2521 (814) 898-1054	Prayer ministry for families affected by hemangiomas. A newsletter is published and an annual picnic is held
Hemangioma Newslines	P.O. Box 38264 Greensboro, NC 27438-8264	Newsletter published. Has a complete list of multi-specialty clinics located in US specializing in vascular birthmarks
Hemangioma Research and Education	43 Soundview Lane New Canaan, CT 06840-2732	Newsletter for patients, families and medical professionals dealing with hemangiomas and vascular malformations
The Hemangioma Support Group	6349 North Commercial Portland, OR 97217 (503) 289-6295	Provides forum for exchange of experiences, medical articles and research advice
About face	P.O. Box 93 Limekiln, PA 19535-0093 (800) 225-3223	Support group for people with facial differences

can be utilized. While doing this therapy, it is important to remind the parent that the eye may have to be held open for the infant to see. These visual stimulation activities should help prevent amblyopia.

In conclusion, parent education and early intervention is fundamental in working with infants who exhibit capillary hemangiomas. It is necessary to educate the parents on the importance of visual stimulation to help prevent amblyopia. Also, parents should understand the need for frequent follow-up to evaluate the child's vision and refractive error. Lastly, parents should be informed that the chances of the hemangioma regressing on its own are very high. While waiting for the regression, the importance of visual stimulation should be stressed. Since the American Optometric Association recommends that children receive their first comprehensive eye and vision examination at 6-12 months of age and since the College of Optometrists in Vision Development has always supported early intervention, the pediatric/functional/behavioral optometrist should be ready to diagnose and manage a wide range of disorders including those whose etiology may be pathological in nature.

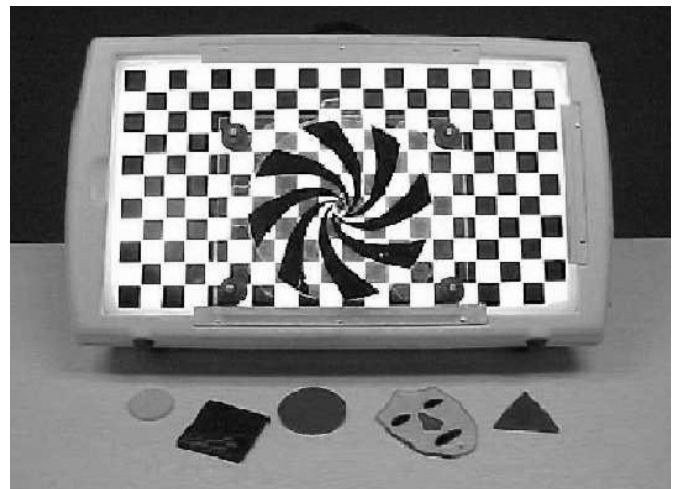


Figure 5. Light Box.

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