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. 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...
often continues over an eight month period. 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.

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. 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 ambylophia 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
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. 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. Monocular congenital cataracts are the leading cause of this type of amblyopia. 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. 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. 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 intraliesional 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>
<thead>
<tr>
<th>Table 1: Differential Diagnosis[^4][^5][^6]</th>
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<tr>
<td><strong>Age of onset</strong></td>
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<tr>
<td>Capillary Hemangioma</td>
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<tr>
<td>Lymphangioma</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
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<tr>
<td>Dermoid cysts</td>
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<tr>
<td>Orbital Cellulitis</td>
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</table>

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

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### Table 2: Treatment Options 2,7,10,11,12,13,14

| 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.

| Intraleisional 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.

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### Table 3: 4 Months of Age vs. 4 Years of Age

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

### Table 4: Support Groups (http://www.birthmark.org/support.html)

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

**References**

16. American Publishing House for the Blind. P.O. Box 6085. Louisville, Kentucky, 40206-0085