Surgical Management, Outcomes and Recurrence Rate of Orbital Lymphangiomas

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Abstract

Objective: Orbital lymphangiomas are rare benign vascular lesions of the orbit. Due to their intimate relationship to the orbital contents gross total resection can be difficult resulting in a relatively high recurrence rate. We sought to review our experience with the surgical management of these lesions.

Methods: A retrospective review of a prospectively maintained database was performed. Demographics, presentation, imaging findings, surgical procedures, clinical and radiographic outcomes and recurrences were reviewed.

Results: Between May 2002 and June 2013, 8 patients (5 male, 3 female) underwent surgical resection for orbital lymphangiomas at our institution. Presenting symptoms included headache, retro orbital pain, eye movement restriction, proptosis, chemosis, diplopia, and visual impairment. The duration of symptoms before treatment ranged from 6 months to 6 years. All patients underwent a modified Orbito Zygomatic (OZ) craniotomy for tumor resection. One patient was lost to follow-up. Of the 7 patients with follow-up 6 experienced improvement in presenting symptoms at a mean of 5.2 years (range 1-23 years). Recurrence was seen in 5 of the 7 patients (71%). Recurrence presented at a mean of 7.2 years (range 1-23 years) after the initial procedure. All of the patients with recurrence were recommended for repeat surgical intervention. Three patients underwent repeat OZ craniotomy, one patient refused surgery and elected for stereotactic radiosurgery and another refused any further intervention. New or worsening cranial nerve deficits were seen post-operatively in 3 of 7 patients (37.5%) with follow-up. All post-operative deficits resolved at last follow-up. No long term complications were identified in this study population.

Conclusion: Orbital lymphangiomas are challenging surgical lesions in which gross total resection is frequently not possible. Subtotal resection is safe and effective for symptomatic relief. The modified OZ approach provides excellent exposure for the surgical management of these lesions. Orbital lymphangiomas have a high rate of recurrence and long-term follow-up is mandatory.

INTRODUCTION

Lymphangiomas are rare, benign, vascular malformations of the head and neck. Although 20% of these lesions are found within the orbit and ocular adnexa, [1-7] they only constitute 0.3% to 4% of all orbital tumors [1,5-7]. The developmental origin of these lesions is not well delineated and an association with intracranial vascular anomalies has been reported [8]. Given the obscure developmental origin of these lesions, it is not surprising that they are referred to by various names in the literature including, hamartomas, orbital venous anomalies, venous lymphatic malformations, congenital venous varices or a distinct anomaly [7].

Orbital lymphangiomas often present with progressive restriction of eye movement, proptosis, retroocular pain and diplopia. Acute severe headache, ocular pain and compressive ocular neuropathy including blindness are usually the result of spontaneous hemorrhage [1,3,4,6,9]. Orbital lymphangiomas interdigitate into the orbital contents with no obvious plane between the lesion and normal structures, making gross total surgical resection challenging [1,3,4,6,9]. We present the surgical experience at the Barrow Neurological Institute (BNI) with the microsurgical treatment of these lesions, focusing on the long-term outcomes and incidence of recurrence with this treatment option.
MATERIAL AND METHODS
Eight patients with a confirmed pathologic diagnosis of orbital lymphangioma were surgically treated and followed at the BNI between May 2002 and June 2013. Four of these patients were initially treated at outside institutions and referred for further evaluation. A retrospective review of prospectively collected data including demographics, clinical features, surgical approaches, intraoperative findings, complications, outcomes and recurrence rates was performed. Computed tomographic (CT) scans, magnetic resonance imaging (MRI) and intraoperative surgical videos were also reviewed when available.

RESULTS
Patients and imaging

Our study population consisted of 8 patients (5 male and 3 female) with an age range from 5 to 64 years (mean 26.8 year) and a pathologic diagnosis of orbital lymphangioma that underwent surgical treatment (Table 1). Presenting symptoms included: headache, retroocular pain and restriction of eye movement, diplopia, blurry vision, chemosis and proptosis. Three patients experienced spontaneous hemorrhage with acute pain and rapidly progressive proptosis. The duration of the symptoms prior to diagnosis ranged from 6 months to 6 years (mean: 2.3 years, median: 1 year). In all 8 cases radiographic findings included in traumaorbital multilobulated lesions, in the intra- and/or extraconal spaces with poorly defined margins, without evidence of invasion into the eyeball or beyond the orbital walls. The orbital masses were mildly heterogeneously enhancing after contrast administration and occasionally included calcifications (Figure 1 and 2). The size of the tumors varied from 2 to 7 centimeters in their largest diameter, including the cystic portion. Multiple thalamic developmental venous anomalies were also identified on imaging in one patient (Figure 2 and 3).

Table 1: Summary of patients with orbital lymphangioma.

<table>
<thead>
<tr>
<th>Pt#</th>
<th>Gender/ Age at Presentation (years)</th>
<th>Presenting Symptoms</th>
<th>Duration of symptoms prior to Dx (years)</th>
<th>Lesion characteristics on MRI (measurements in axial plane)</th>
<th>Initial Procedure</th>
<th>Follow-up (years)/ recurrence</th>
<th>Post-recurrence Intervention</th>
<th>Post-operative Complications</th>
<th>Outcome/ follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F/5</td>
<td>Retroocular pain, proptosis</td>
<td>1</td>
<td>Right 2x2 cms lateral wall orbital mass</td>
<td>OZ craniotomy and tumors resection</td>
<td>1/yes</td>
<td>None</td>
<td>Excellent /2 months</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>M/6</td>
<td>Chemosis, proptosis</td>
<td>5</td>
<td>Left 2x2 cms orbital cystic mass</td>
<td>OZ craniotomy and partial resection</td>
<td>Lost follow-up</td>
<td>N/A</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>F/12</td>
<td>Retroocular pain, blurry vision progress to blindness</td>
<td>4</td>
<td>Left 4x3 cms orbital cystic mass</td>
<td>Orbitotomy and partial resection</td>
<td>23/yes</td>
<td>None</td>
<td>Good / 2 months</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>M/18</td>
<td>Diplopia, eye movement restriction</td>
<td>1</td>
<td>Right 3x3 cms orbital cystic mass with fluid levels</td>
<td>OZ craniotomy and partial resection</td>
<td>5/yes</td>
<td>None</td>
<td>Poor / 5 years (Retroocular pain, diplopia, decrease vision, lost of mobility)</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>M/23</td>
<td>Retroocular pain, blindness, proptosis</td>
<td>22</td>
<td>Right 3x3 cms orbital cystic mass with fluid levels</td>
<td>OZ craniotomy and near gross total resection</td>
<td>1/no</td>
<td>None</td>
<td>Excellent /1 year</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>F/34</td>
<td>Chemosis, retroocular pain, diplopia</td>
<td>5</td>
<td>Left 3x2 cms lateral wall orbital mass</td>
<td>OZ craniotomy and near gross total resection</td>
<td>1/no</td>
<td>None</td>
<td>Temporary Oculomotor Palsy / Good /1 year</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>M/53</td>
<td>Retroocular pain, diplopia</td>
<td>1</td>
<td>Left medial wall 2x1 cms orbital apex cystic mass</td>
<td>Transphenoidal near gross total resection</td>
<td>2/yes</td>
<td>None</td>
<td>Excellent /2 years</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>M/64</td>
<td>Chemosis, H/A, proptosis, blindness</td>
<td>6</td>
<td>Right 6x3 cms orbital cystic mass with fluid levels causing severe mass effect</td>
<td>Orbitotomy, cyst drainage and biopsy</td>
<td>4/yes</td>
<td>None</td>
<td>Excellent /3 years</td>
<td></td>
</tr>
</tbody>
</table>

*Patients treated initially at outside institutions.
This approach provides a superior-lateral exposure of the orbit. After the craniotomy was complete the roof and lateral wall of the orbit were removed using a diamond burr. Laterally the bony removal was carried posteriorly to the superior orbital fissure. Superiorly the decompression was carried posteriorly to the optic canal. This provides a generous orbital decompression and allows for a wide opening of the periorbita facilitating tumor resection and visualization of orbital contents. Orbital reconstruction was not performed on any of the patients treated in this series. Of the patients treated initially at outside institutions one underwent a transphenoidal approach, two were treated with an anterior orbitotomy and one with an orbitozygomatic approach (Table 1).

**Follow-up and outcome**

Clinical and radiographic follow-up were available on all but one patient. The mean follow-up was 5.2 years (range from 1 to 23 years) and the median was 2 years. All patients experienced improvement of their presenting symptoms after surgery with the exception of patient 5 who presented with blindness and never recovered vision. Symptomatic recurrence was seen in 5 of 7 patients (71%). Of note the 2 patients without recurrence each only have 1 year of follow-up. Post-operatively 3 of 8

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**Figure 1** Orbital lymphangioma. A and B, axial and coronal CT of intraorbital extraconal isodense lesion with calcifications. C and D, post-gadolinium T1-weighted (A) and T2-weighted axial MRI images of the orbit showing a diffuse enhancing cystic mass (C) at the medial orbital wall with mass effect over the orbital content. E, T2-weighted coronal MRI showing a cystic mass at the medial wall of the orbit displacing orbital contents laterally. F, post-contrast T1-weighted sagittal MRI image showing the intraorbital mass displacing the orbital content anteriorly.

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**Figure 2** Orbital lymphangioma. A and B, post-gadolinium T1-weighted MRI axial (A) and coronal (B) images showing a large intraorbital cystic mass causing severe mass effect on the orbital content and proptosis. C, T2-weighted coronal MRI image showing the cystic mass. D, post-gadolinium T1-weighted sagittal MRI image showing the cystic mass and the intracranial thalamic multiple developmental venous anomaly (arrow).

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**Figure 3** A and B, post-gadolinium T1-weighted and gradient ECHO coronal MRI images showing the multiple developmental venous anomalies (DVA) (arrow) in the left thalamus. C, axial view of the multiple DVAs. D, post-enhancing T1-weighted MRI image showing the abnormally enlarge left vein of Rosenthal (arrow heads).

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**Video 1** A left orbitozygomatic craniotomy is performed with care not to violate the frontal or temporal dura. The periorbita is opened laterally and the intraorbital lymphangioma is debulked. The tumor was relatively vascular with poorly defined margins and was thus unable to be resected completely without risking visual complications. Remaining tumor is coagulated and meticulous hemostasis obtained. Intraoperative navigation was used.
patients (37.5%) had worsening or new cranial nerve deficits, 2 oculomotor and 1 abducens nerve palsy. All three of these patients had complete recovery of function at last follow up. No long-term complications were identified in this study population.

**DISCUSSION**

Orbital lymphangiomas are benign vascular lesions that usually present during early childhood [2,9]. In the present series, four patients were 18 years of age or younger at presentation, while the remaining patients’ ages ranged from 23 to 64 years [10]. Patients with orbital lymphangiomas typically present with ocular symptoms and are evaluated by ophthalmologists and/or primary care physicians. Given the non-specific symptoms and the slow growth of these lesions diagnosis can be delayed for months or years after symptomatic onset. Our series had durations of symptoms prior to diagnosis ranging from 6 months to 6 years (mean: 2.3 years). In one case (Patient 5), the diagnosis was not made until 22 years after the onset of symptoms. The large majority of patients are asymptomatic at the time of presentation. Rarely patients are diagnosed incidentally and frequently radiographic recurrence will be identified in an asymptomatic patient. Differing opinions exist in the current literature regarding whether or not to intervene when a patient is asymptomatic [4,11]. Based on our experience there is no indication for microsurgical treatment unless the patient is symptomatic. Given the benign biology of these tumors, the extremely high likelihood of recurrence and the risk of perioperative morbidity it is difficult to justify early intervention.

The International Orbital Society had classified orbital vascular malformations into 3 different types. Type 1 with no flow, type 2 with venous flow and type 3 with arterial flow. These different types are not always distinct and there can be overlap. Orbital lymphangiomas are considered type 1 vascular malformations, having minimal or no connection with the vascular system and minimal internal flow [3]. Other authors classify orbital lymphangiomas as hamartomatous lesions rather than vascular malformations given the lack of obvious abnormal connection to the vasculature [12]. Of note, none of the patients in our series underwent catheter angiography prior to surgery or during the follow-up period.

Histologically, lymphangiomas are diffuse unencapsulated masses that interdigitate with orbital tissue and consist of thin-walled vascular channels, with interrupted basement membrane, attenuated endothelium, loss of fibrous stroma and clusters of lymphocytes. Occasionally they can be mistaken for cavernous hemangiomas, but the diagnosis can be clarified by the absence of smooth muscle cells within the wall of the vessels, and the presence of lymph. As with all veno-lymphatic lesions, lymphangiomas may enlarge during an episode of infection because of lymphoid proliferation.

If the diagnosis of orbital lymphangioma is suspected, the work-up should include CT and MRI with and without contrast of the orbit and brain. Computed tomography can aid in planning surgical resection by delineating the status of the orbital wall, which could be thinned or remodeled by the malformation. Calculifications within these lesions are common and can be appreciated on CT as well. The common MRI findings are a mildly enhancing cystic mass that infiltrates the orbital content but respects the eyeball. Cysts can contain fluid levels that correspond to hemorrhages of different ages. MRI of the brain can demonstrate intracranial vascular lesions that are commonly associated with orbital lymphangiomas [13]. One of our patients had multiple thalamic DVAs identified on pre-operative imaging (Figure 3). None of the other patients in our series had associated intracranial vascular malformations.

**Optimal timing for surgical resection**

The optimal treatment regimen for orbital lymphangioma is not well defined [11,14-17]. However, the majority of authors recommend intervention when there is a symptomatic presentation including visual impairment, repeated hemorrhagic episodes and progressive proptosis [2,4,9]. Based on our experience we agree that surgical intervention is indicated only in the setting of symptomatic presentation. The primary goal of surgery is to evacuate the cystic component, debulk the lesion and decompress the orbit. Complete resection is ideal, however these lesions typically interdigitate with the orbital contents making a gross total resection extremely challenging. In the current series, all patients had subtotal resection and 6 of 7 patients had resolution of their symptoms. Based on the favorable prognosis after debulking, despite waiting for symptomatic presentation, there is little justification for early intervention with such a high recurrence rate.

**Surgical approach**

Ophthalmologists and neurosurgeons treat orbital lymphangiomas. In general, an ophthalmologic approach is through the anterior or lateral orbit. In a recent series published by neurosurgeons [4], a transcranial approach with superior-lateral orbitotomy was used. This approach provided good exposure of the orbital contents without significant retraction or manipulation of orbital structures. In the current series, we used a modified OZ craniotomy in all patients. Similar to the prior mentioned publication we believe that this approach offers an excellent exposure of the superior and lateral orbital wall and also reduces the need for retraction and manipulation of the orbital contents. Removal of the superior-lateral orbital wall also decompresses the orbit in case of future recurrence. Of interest, none of our patients have experienced enophthalmos during the follow-up period despite wide orbital decompression. Intraoperative neuronavigation is used routinely for these cases to minimize unnecessary dissection within the orbit (Video 1). Some authors have suggested pre-operative intralesional injection with synthetic polymers may facilitate surgical resection by making the lesion assume a more firm texture [18]. This technique was not used for any of the patients included in this study.

**Recurrences rate**

Orbital lymphangiomas are poorly defined lesions making gross total resection extremely difficult resulting in high recurrence rates. Tunc [6] reported a series of 26 patients with a 58% recurrence rate during a mean follow-up of 9.2 years. They had a mean interval between treatment and recurrence of 3.4 years. Gündüz [15] reported an 11% recurrence rate with a mean follow-up of 2.5 years. The mean interval between treatment and recurrence was 16 months. Harris [16] reported that 12 of 23 (52%) surgically treated patients had major hemorrhagic events ranging from 4 days to 12 years post-operatively. Rather uniquely, Maciel Simas [4] reported 5 patients who underwent subtotal surgical resection and none had any significant
reurrence during mean follow-up of 3.2 years. In the present series, of the 4 patients originally treated at the BNI, 1 patient (Patient 6) did not have evidence of recurrence at 1 year follow-up, 1 (Patient 2) was lost to follow up and the remaining 2 (Patient 1 and 4) experienced recurrence at 1 and 5 years of follow-up. Four patients were originally treated at outside institutions and referred to the BNI after they recurred. One patient (Patient 3) underwent an anterior orbitotomy with subtotal resection and then presented with acute blindness 23 years later. He underwent surgical treatment with no improvement in his visual symptoms. A second patient (Patient 5) underwent an OZ craniotomy and had no evidence of recurrence at 1 year. The remaining two patients (Patient 7 and 8) had recurrences at 2 and 4 years. Patient 7 was previously treated with a transsphenoidal approach for subtotal tumor resection and had his recurrence surgically managed. Patient 8 had a prior orbitotomy and refused microsurgery for his recurrence. Instead, the patient’s recurrent tumor was treated with stereotactic radiosurgery and his tumor is unchanged at 3 years post-radiation (Table 1).

Alternative treatments

One alternative to surgical resection is the intralesional injection of a sclerosing agent. OK-432 (Picibanil, Shuhei Ogita, Fund, Japan) is a freeze-dried biological product prepared from the Su strain of *Streptococcus pyogenes* (group A). The mechanism of action is induction of macrophages and neutrophils with production of interleukin-6 and tumor necrosis factor causing increased membrane permeability and ultimately resulting in contraction of the lesion. Results on predominantly macrocystic lesions have shown a 60% reduction in the size of the lesion in 86% of the treated patients [18,19]. Treatment protocol consisted of four intraleional injections at 6 to 8 week intervals. Other sclerosant agents are sodium tetradecyl sulphate and sodium morrhuate [18]. Radiation therapy has not been proven to be effective for orbital lymphangiomas [20]. One patient (Patient 8) received radiosurgery and has not shown evidence of recurrence at 3 year follow-up. Our experience with radiosurgery is limited to one patient and no definitive conclusions can be drawn.

CONCLUSION

Orbital lymphangiomas are rare, benign, diffuse vascular lesions. Gross total resection is frequently not possible. Subtotal resection has good functional results with low risk of permanent neurological deficits. The orbitozygomatic approach provides an excellent exposure for the surgical management of these lesions. Orbital lymphangiomas have a high recurrence rate and clinical and radiographic long-term follow-up is necessary in all patients. Promising alternative treatments with sclerosing agents require further investigation.

REFERENCES