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Clinical Study

Surgical management, outcomes, and recurrence rate of orbital lymphangiomas

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ABSTRACT

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. Between May 2002 and June 2013, eight patients (five male, three female) underwent surgical resection for orbital lymphangiomas at our institution. Of the seven patients with follow-up, six experienced improvement in presenting symptoms at a mean of 5.3 years (range 1–23 years). Recurrence was seen in five of the seven patients (71.4%). Recurrence developed at a mean of 7.2 years (range 1–23 years) after the initial procedure. New or worsening cranial nerve deficits were seen postoperatively in three of eight patients (37.5%) with follow-up. All postoperative deficits were resolved at last follow-up. Orbital lymphangiomas are challenging surgical lesions in which gross total resection is frequently not possible. Subtotal resection is safe and effective for symptomatic relief.

1. 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 constitute only 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 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.

2. Material and methods

Eight patients with a confirmed pathologic diagnosis of orbital lymphangioma were surgically treated and followed at BNI between May 2002 and June 2013. Four of these patients were initially treated at outside institutions and were referred to BNI for further evaluation. A retrospective review was performed of prospectively collected data, including demographic details, clinical features, surgical approaches, intraoperative findings, complications, outcomes, and recurrence rates. CT scans, MRI, and intraoperative surgical videos were also reviewed when available.

3. Results

3.1. Patients and imaging

Our study population consisted of eight patients (five male and three female) with an age range from 5 to 64 years (mean 26.9 years) and a pathologic diagnosis of orbital lymphangioma who underwent surgical treatment (Table 1). Presenting symptoms included headache, retroocular pain, 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 22 years (mean 5.1 years, median 2.5 years). In all eight patients, radiographic findings included intraorbital multilobulated lesions, in the intraconal and/or extraconal spaces with poorly defined margins, without

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Table 1
Summary of patients with orbital lymphangioma

Patient	Sex/Age (years)	Presenting symptoms	Duration of symptoms prior to diagnosis (years)	Lesion characteristics on MRI (measurements in axial plane)	Initial procedure	Follow-up (years)/ Recurrence	Post-recurrence intervention	Postoperative complications	Outcome/Follow-up
1	F/5	Retroocular pain, proptosis	1	Right 2 × 2 cm lateral wall orbital mass	OZ craniotomy and resection	1/Yes	Redo OZ craniotomy and resection	Temporary oculomotor palsy	Excellent/2 months
2	M/6	Chemosis, proptosis	0.5	Left 2 × 2 cm orbital cystic mass	OZ craniotomy and partial resection	Lost to follow-up	N/A	None	N/A
3*	F/12	Retroocular pain, blurry vision progressing to blindness	4	Left 4 × 3 cm orbital cystic mass	Orbitotomy and partial resection	23/Yes	OZ craniotomy and resection	None	Good/2 months
4	M/18	Diplopia, eye movement restriction	1	Right 3 × 3 cm orbital cystic mass with fluid levels	OZ craniotomy and partial resection	5/Yes	None (patient refused further treatment)	None	Poor/5 years (retroocular pain, diplopia, decreased vision, loss of mobility)
5*	M/23	Retroocular pain, blindness, proptosis	22	Right 3 × 3 cm orbital cystic mass with fluid levels	OZ craniotomy and neargross total resection	1/No	–	None	Excellent/1 year
6	F/34	Chemosis, retroocular pain, diplopia	5	Left 3 × 2 cm lateral wall orbital mass	OZ craniotomy and neargross total resection	1/No	–	Temporary oculomotor palsy	Good/1 year
7*	M/53	Retroocular pain, diplopia	1	Left medial wall 2 × 1 cm orbital apex cystic mass	Transsphenoidal neargross total resection	2/Yes	OZ craniotomy and gross total resection	Temporary abducens palsy	Excellent/2 years
8*	M/64	Chemosis, H/A, proptosis, blindness	6	Right 6 × 3 cm orbital cystic mass with fluid levels causing severe mass effect	Orbitotomy, cyst drainage and biopsy	4/Yes	Radiosurgery (Gamma Knife[a])	None	Excellent/3 years

* Patients initially treated at outside institutions.
[a] Elekta AB, Stockholm, Sweden.
F = female, H/A = headache, M = male, N/A = not available, OZ = orbitozygomatic.

evidence of invasion into the globe or beyond the orbital walls. The orbital masses were mildly heterogeneously enhancing after contrast administration and occasionally included calcifications (Fig. 1, 2). The size of the tumors varied from 2 to 6 centimeters in their largest diameter, including the cystic portion. Multiple thalamic developmental venous anomalies were also identified on imaging in one patient (Fig. 2, 3).

3.2. Surgical management

A modified orbitozygomatic craniotomy was the approach of choice for all four of the patients initially treated at BNI as well as those patients with recurrence treated microsurgically (Supp. Video 1). 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 approach provides a generous orbital decompression and allows for a wide opening of the periorbita to facilitate tumor resection and visualization of orbital contents. Orbital reconstruction was not performed on any of the patients treated in this series. Of the four patients treated initially at outside institutions, one underwent a transsphenoidal approach, two were treated with an anterior orbitotomy, and one was treated with an orbitozygomatic approach (Table 1).

3.3. Follow-up and outcome

Clinical and radiographic follow-up were available for seven of the eight patients. The mean follow-up was 5.3 years (range 1 to 23 years) and the median follow-up was 2 years. All patients experienced improvement of their presenting symptoms after the initial surgery, with the exception of Patient 5 who presented with blindness and never recovered vision. Symptomatic recurrence was seen in five of seven patients (71.4%). Of note, the two patients without recurrence each had only 1 year of follow-up. Postoperatively, three of eight patients (37.5%) had worsening or new cranial nerve deficits; two had oculomotor nerve palsy and one had 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.

4. 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, and four patients ranged in age 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 nonspecific symptoms and the slow growth of these lesions, diagnosis can be delayed for months to years after symptomatic onset. Our series had durations of symptoms prior to diagnosis ranging from 6 months to 22 years (mean 5.1 years). In one case (Patient 5), the diagnosis was not made until 22 years after the onset of symptoms. The majority of patients with orbital lymphangiomas are symptomatic 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]. Our