Cavernous Hemangioma of the Orbit: 42 Patients

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Background/Purpose: To analyze clinical features and treatment results of 42 patients with cavernous hemangioma of orbit.

Design: A retrospective chart review of cases.

Methods: Between 1976 and 2011, 42 patients with pathologically proven cavernous hemangioma of the orbit were referred to, and treated by, the authors. The clinical features and treatment outcomes were analyzed.

Results: The retrospective study included 42 cases of symptomatic tumors that were completely removed by means of anterior orbitotomy (8/42, 19%), lateral orbitotomy (30/42, 71%), and transcranial approach (4/42, 10%). There were more female cases than male cases (29:13). Ages ranged from 17 years to 71 years (mean 46.2 years ± 6.8 years). Most of the tumors (32/42, 76%) were located in the muscle cone. The left orbit was affected in 18 cases, and the right orbit in 24 cases, no case was bilaterally involved. Painless, gradually progressive proptosis (exophthalmos) and visual disturbance were the main clinical signs. Of the cases, 90% (38/42) could be accurately diagnosed preoperatively based on ultrasound scan and computed tomography/magnetic resonance imaging examination. Improvement (or no change) of visual function (35 cases, 83%) was achieved in the 42 cases who received surgical treatment. No recurrence was noted during a 6 month to 136 month follow-up period.

Conclusion: Nearly 90% of 42 patients with cavernous hemangioma of orbit could be correctly diagnosed preoperatively. The location of tumor determined the choice of surgical approach. Orbitotomies with careful dissection and cryoprobe extraction of the tumor have been successfully used in 32 cases. Among the 42 patients who received surgical treatment, good outcome of visual function (83%) was achieved. All patients were regularly followed up at least 6 months (range, 6–136 months; mean 16.4 months) and no recurrence was noted.

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1. Introduction

Cavernous hemangioma is the most frequent primary orbital tumor seen in adults. It is a benign lesion but may compromise optic nerve function. With neuroradiation image studies, the tumors have become more detectable. Asymptomatic lesions can only be followed up clinically but for symptomatic tumors the surgical excision of symptomatic cavernous hemangioma seems to be the treatment of choice. The authors reported their own experiences regarding 42 patients with cavernous hemangioma of the orbit, and compared the clinical features and treatment outcomes with other studies.

2. Patients and methods

During 35 years (from 1976 to 2011), a total of 42 patients (13 male, 29 female) with orbital cavernous hemangioma were referred to, and treated by, the authors (41 cases treated by WMH). Patients’ ages ranged from 17 years to 71 years (mean 46.2 years ± 6.8 years). Surgical removal and histological confirmation were performed on 42 patients.

The ophthalmic examinations included visual acuity (VA) measurement with Snellen charts at a distance of 6 meters, motility of eye, pupillary reflex, and fundus examination with direct ophthalmoscope. The protocol was performed preoperatively, one week after the surgery, one month and every 6 months following. Most of the patients underwent orbital computed tomography (CT) (38 patients), 12 underwent magnetic resonance imaging (MRI) and
4 patients underwent orbital angiography only (before 1978, due to unavailability of CT machines). Orbital echography utilizing A- and B-mode ultrasound (10 MHz) was performed on 12 patients in an attempt to corroborate the diagnosis of tumors and the location of the lesion.

Indications for surgical therapy in patients with clinical symptoms related to the intraorbital lesion included: (1) visual impairment (Snellen chart vision less than 6/7.5); (2) progressive and disfiguring unilateral proptosis (1.5 mm more protruded than fellow eye); (3) double vision; (4) retrobulbar pain clearly related to the tumor; and (5) choroid fold. Location of tumor determined the choice of surgical approach. According to the CT (or MRI) findings, if the tumor was located in the anterior third compartment of orbit, we chose anterior orbitotomy. If the tumor was located in the posterior two third space of the orbit and it was lateral to the optic nerve, lateral orbitotomy was chosen. If the tumor was located superomedial to the optic nerve or near the orbital apex, then an intracranial approach (pterional approach or fronto-orbitozygomatic approach performed by one of the authors (WMH) combined with neurosurgeon) was chosen.

All surgery was performed under general anesthesia. The surgical approach used most frequently by the author (WMH) was the lateral approach orbitotomy with blunt dissection and cryoprobe extraction of the tumor. Histopathological examinations confirmed the final diagnosis. VA improvement is defined as a postoperative VA of one line (by Snellen chart) better than preoperative VA. All patients were regularly followed up according to the protocol, at least for 6 months (range, 6–136 months; mean 16.4 ± 5.2 months).

3. Results

Clinically, the patients presented with an acute or subacute onset and a history averaging 9 months (ranging from 3 months to 3 years). Presenting symptoms included eyeball protrusion, local pain, visual impairment, double vision, eyelid swelling, and headache (Table 1). Presenting signs included exophthalmos, visual impairment, choroidal folding, disc edema, and optic atrophy (Table 2). None of the patients had cutaneous vascular malformations or a family history of cerebral or orbital hemangioma. The left orbit was affected in 18 cases and the right orbit in 24 cases. No case was bilaterally involved. Most of the tumors (32/42, 76%) were situated in the muscle cone and in the proximity of orbital apex most often superolateral to the optic nerve. Ten cases were situated extraconally. Among 32 intraconal tumors, 22 were located lateral to the optic nerve (Figure 1), four were inferior to the optic nerve, three were medial to the optic nerve (Figure 2) and three cases were near the orbital apex.

Most of the tumors (30 cases) were removed through lateral orbitotomy with cryoprobe extraction of the tumor (Figure 3). Anterior orbitotomy with blunt dissection to remove tumor were performed in six cases, dissection of tumor with cryoprobe extraction were performed in two cases. Four cases received surgical treatment through craniotomy (neurosurgeon and eye specialist combined) due to deeply situated tumors (near orbital apex). The extracted tumor size ranged from 15 mm × 12 mm × 10 mm to 30 mm × 30 mm × 20 mm (Figure 4). On histopathological study, the engorged vascular channels, which are tightly knit and separated by fibrous septae, can be seen. These channels are lined with a single layer of endothelial cells. (Figures 5 and 6)

The results of the operations 6 months after surgery (Table 3) showed: VA improvement (83%), no change of vision (15%), exophthalmos decreased (88%), and individual satisfaction (86%). Surgical complications (Table 4) showed: loss of vision in one case (due to central retinal artery occlusion), impaired vision (5%), diploia (5%), esotropia (5%), and pupil dilation (7%). Postoperative follow up (range, 6–136 months) revealed there was no recurrence of the lesions.

| Table 1 Presenting symptoms among 42 patients with orbital cavernous hemangioma |
|----------------------------------------|-----------------|
| Symptoms                              | No. of patients (%) |
| Eyeball protrusion                    | 38 (90%)         |
| Visual impairment                     | 27 (65%)         |
| Double vision                         | 8 (20%)          |
| Local pain                            | 7 (18%)          |
| Headache                              | 5 (12%)          |
| Eyelid swelling                       | 2 (5%)           |

| Table 2 Presenting signs among 42 patients with orbital cavernous hemangioma |
|----------------------------------------|-----------------|
| Signs                                  | No. of patients (%) |
| Exophthalmos                           | 40 (95%) 1.4 mm ± 0.9 mm |
| Visual impairment                      | 27 (64%) VA: 6/7.5; HM (mean 6/12) |
| Choroid folding                        | 14 (33%)        |
| Optic disc edema                       | 7 (17%)         |
| Optic field defects                    | 5 (12%)         |
| Optic atrophy                          | 2 (4%)          |

HM – hand motion; VA – visual acuity.
4. Discussion

Cavernous hemangiomas are the most common intraorbital tumors found in adults. One large study reported an incidence of 4.3% among orbital neoplasms.1 In Taiwan, a hospital-based study demonstrated an incidence of 11% among 200 orbital tumors.9 Those benign vascular lesions grow exceedingly slowly and therefore can present as a painless progressively proptotic eye. Most of the tumors are unilateral; bilateral or multiple lesions have been reported but they are rare.13,14 Most patients are female. Harris and Jakobiec found a 7:3 occurrence ratio of female to male,2,3 whereas Henderson reported an almost equal ratio (8:7) of female to male.1

In our experience, the incidence of cavernous hemangiomas (pathology proved 42 cases) is 9.1% among orbital neoplasms (authors’ series, from 1976 to 2011, total of 462 cases of orbital tumors). All of our cases are unilateral lesions. The female to male ratio (29:13) is almost 2:1, which is similar to Harris and Jakobiec’s result.2,3

The diseases are usually diagnosed during the 3rd to 5th decades of life. In Harris and Jakobiec’s study the average age is 42 years,2,3 and in McNab and Wright’s study the average age is 42.1 years.6 In our study, patients’ age ranged from 17 years to 71 years (mean 46.2 years ± 6.8 years).

Symptoms and signs caused by orbital cavernous hemangiomas were due to mass effect, which can increase intraorbital volume. Although cavernous hemangiomas are histologically benign, their compression on intraorbital or adjacent structures makes them considered anatomically or positionally malignant. Blurred vision, visual field defects, restricted extraocular motility, diplopia, pupillary dysfunction, choroidal folds, and papilledema are common signs.1 A previous study showed that the most common symptom is exophthalmos (95%), followed by impairment of VA (40%), local pain (25%), visual field defects (15%), diplopia (15%), and chronic headache (10%).10 In addition, lagophthalmos can result in exposure keratopathy, keratitis, and corneal perforation.2−5 Optic atrophy is also noted occasionally.1−9

In our series, the presenting symptoms include eyeball protrusion (90%), visual impairment (65%), double vision (20%), local pain (18%), and headache (12%). Exophthalmos (4.7 mm ± 1.6 mm) is the most common sign followed by visual impairment, which is similar to other studies.6−10 Optic nerve atrophies were found in two of 42 patients (4%), and the VA was only at hand motion level.

Figure 3 Cavernous hemangioma was extracted with a cryoprobe.

Figure 4 Gross view of cavernous hemangioma.

Figure 5 The histopathological picture of cavernous hemangioma. Grossly engorged vascular channels, which are tightly knit and separated by fibrous septae. (H&E stain, × 10).

Figure 6 In cavernous hemangioma, the vascular channels are lined by a single layer of endothelial cells. (H&E stain, × 40).
Routine CT scanning or MRI may reveal the orbital lesion. On a CT image, an oval or round shaped homogenous lesion with a well-defined margin may be detected and with contrast the tumor is enhanced. However, CT alone does not allow one to make a definitive diagnosis. For example, hemangiopericytoma and neurilemmoma have findings similar to hemangioma on a CT image. MRI is more sensitive and specific for the diagnosis but still should not be solely relied upon. An ultrasound study (A-scan) can find a uniform high-echogenicity mass and the reflections are secondary to the septae within the lesion.

In our study, most patients underwent orbital CT (38 patients), 12 patients underwent MRI and four underwent orbital angiography only (before 1978, due to unavailability of CT machines). Orbital echography utilizing A- and B-mode ultrasound (10 MHz) was performed on 12 patients. According to our experience, CT with contrast is enough for image evaluation. Although MRI is better for understanding the nature of the orbital mass, it still cannot allow an ophthalmologist to make a definite diagnosis. The final diagnosis still needs histopathologic approval after surgical treatment. Therefore, most of our cases (90%) underwent orbital CT but not MRI.

In histopathologic study, a cavernous hemangioma is well encapsulated and reveals a typical picture of engorged vascular channels, which are separated by fibrous septae. Those channels are lined with a single layer of endothelial cells (Figures 5 and 6). Compared with other well-encapsulated orbital tumors, hemangiopericytoma reveals “staghorn appearance vessels”, and neurilemmoma is filled with spindle-shaped Schwann cells arranged in interlacing fascicles.

Because of their benign nature, most orbital hemangiomas require no immediate intervention. If surgical treatment is indicated, the majority of patients with the lesions can be managed primarily by ophthalmologists with experience in orbital surgery.

Most orbital hemangiomas are found between the optic nerve and extraocular muscles within the intraconal space. Therefore the approach to the lesion depends on the location of the tumor within the orbit. A lateral orbitotomy is a typical approach but lesions located within the medial aspect of the orbit will best be approached through an upper eyelid or a transcarnuncular-based medial orbitotomy. Only small tumors located laterally or medially to the eyeball can be excised by a tranconjunctival approach, which is a cosmetically and functionally very satisfying technique. In our experience, most of the tumors (30 cases) were removed through a lateral approach orbitotomy with cryoprobe extraction, and anterior orbitotomy with/without cryoprobe extraction in eight cases. Four cases received surgical treatment through craniotomy due to deeply situated tumors (superomedial to optic nerve or near the orbital apex).

The cryoprobe, thought to be an ideal tool for hemangioma extirpation, allows for retraction of well-circumscribed tumors, and is reported to have good results in reducing intraoperative capsular rupture and bleeding. A disadvantage of the method is that adjacent structures may also be frozen, especially deep orbital tumors. In most of our cases, we used a cryoprobe as a tool to remove the tumors, and it seems to be an easy and safe method. In some reports, use of a carbon dioxide laser or neodymium-doped yttrium aluminum garnet laser is another modality for tumor removal but the authors have no such experience.

For a minimally invasive treatment and approach, some make use of the gamma knife, some studies showed the benefits of endoscopic endonasal removal of hemangioma of the orbital apex. However, we do not have experience of these two approaches.

Concerning postoperative complications, signs secondary to cranial nerve injury, such as palpebral ptosis, impairment of extraocular excursions (with or without diplopia), pupil dilation (mydriasis) and visual disturbances had been reported. In our series, the postoperative VA were worse, which is thought to be the most damaging postoperative complication, was reported in 3% to 32% of patients in previous studies. In our series, the postoperative VA were worse in three cases (7%). One of them even lost vision due to central retinal artery occlusion, which is believed to be caused by heavy intraoperative cauterization for hemostasis. The other two cases were due to the location of the tumor, which involved the orbital apex. Otherwise, diplopia (5%), esotropia (5%), and pupil dilation (3 cases, 7%) were also found in our cases. In all three cases with the complication of pupil dilation, the tumors were located temporal to the optic nerve and near the ciliary ganglion.

Concerning the prognosis, in the presence of visual deterioration clearly related to the tumor it was recommended that the tumor be removed by surgery, whereas a small lesion which solely produces mild protrusion of the eyeball could be assessed by observation. If surgical treatment is necessary, excellent prognosis and good results with very low morbidity rate were estimated when the operations were performed by experienced surgeons. Due to the benign nature of the tumor, there is no recurrence following excision or risk of malignant change. In our study, all 42 patients received surgical interventions, and none of them had recurrence or malignant transformation.

In conclusion, nearly 90% of patients with cavernous hemangioma could be correctly diagnosed preoperatively. The surgical procedure of anterior orbitotomy or lateral approach with cryoprobe extraction of tumor can be successfully used with less surgical complication. The transcranial approach may be considered for larger lesions superior and medial to the optic nerve, especially if they involve the orbital apex. Among the 42 patients who received surgical treatment, good outcome of visual function was achieved in 35 individuals (83%). No recurrence was noted during a 6 month to 136 month follow-up period.

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