Orbital Venous Malformations

Current Multidisciplinary Treatment Approach

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Objective: To evaluate the clinical, radiological, and histopathological features, treatment, and outcome of a series of orbital venous flow malformations (OVMs) with the aim of delineating a more systematic approach for treatment.

Methods: A 38-year retrospective review of 22 patients with OVMs followed up at 1 institute.

Results: Eighteen of 22 patients (13 women and 9 men) showed clinical or radiological evidence of distensibility. The mean age at the initial manifestation was 28.3 and 50.7 years in patients with distensible and nondistensible OVMs, respectively. Eight patients (36.3%) had deep orbital lesions, 6 (27.3%) had superficial orbital lesions, and 8 (36.3%) had combined orbital lesions. All 3 patients with deep nondistensible OVMs had a sudden onset of proptosis and pain or diplopia secondary to thrombosis or hemorrhage. Seventeen patients required treatment. All 4 nondistensible lesions were treated by surgical excision. A variety of techniques were used to treat distensible OVMs including carbon dioxide laser ablation, percutaneous alcohol sclerotherapy, or embolization with Guglielmi detachable coils after surgical exposure and surgical excision. Of the 14 patients with follow-up, 8 had complete resolution of the signs and symptoms and 6 patients showed marked improvement following surgery. Mean follow-up was 57.8 months.

Conclusion: If intervention is indicated, less invasive methods such as carbon dioxide laser ablation and percutaneous alcohol sclerotherapy for superficial and combined orbital lesions and endovascular treatment by Guglielmi detachable coil embolization for deep orbital lesions should be considered before proceeding with more invasive procedures.

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Understanding of orbital vascular malformation (OVM) has been difficult owing to the confusion in correctly classifying these lesions and in providing treatment on a rational basis. Wright and colleagues suggested that segregation of OVMs as lymphangiomas and orbital varices is artificial and incorrect and their origin, clinical manifestations, and treatment must be considered as a whole. Others resist the unification of lymphangiomas and orbital varices under the heading of orbital venous anomalies because separation of the conditions along hemodynamic relationships is consistent with their clinical, pathophysiological, and diagnostic features and it also influences therapeutic decisions and surgical approaches. In 1999, a consensus statement on terminology of the OVMs was published by the members of the Orbital Society. According to the new terminology, lymphangiomas were classified as no flow malformations and primary varices, including the mixed lesions with venous no flow components, were classified as orbital venous flow malformations.

This article concentrates on the OVMs as classified by the Orbital Society. This is the category, which includes both distensible OVMs (with the evidence of connection to the venous system either shown clinically or radiologically by distensibility with increased venous pressure) and nondistensible OVMs.

Distensible OVMs are a part of a spectrum of vascular malformations involving periorbital skin, conjunctiva, and/or extraorbital sites such as the central nervous system, face, and nasal sinuses, or a combination of these sites. The coexistence of these vascular malformations at different sites is believed to represent aberrations of vasculogenesis occurring at the same stage of embryonic development. Our objective in this study was to describe the clinical, radiological, and histopathological features, treatment, and out-
come of a series of OVMs with the aim of delineating a more systematic approach for treatment.

METHODS

This study is a retrospective review of 22 consecutive patients having a diagnosis of OVMs who were followed up at our clinic from 1964 to 2002. All except 3 patients were treated and followed up by the same surgeon (M.B.). These 3 patients required endovascular treatment that was done as combined cases with the interventional neuroradiologist (M.E.M.). Our classification of OVMs is listed in Table 1, which was derived from the consensus statement on terminology published by the Orbital Society.1 No flow malformations (lymphangiomas) and arterial flow malformations were excluded from this study. Distensibility was considered present when there was clinical or radiological evidence of transient expansion with increased venous pressure associated with dependent positioning, the Val-salva maneuver, and coughing or by demonstration of distention with increased venous pressure by ultrasonography or computed tomography (CT).2 Diagnosis in all 4 nondistensible OVMs was confirmed histopathologically. Venogram was not performed in these nondistensible lesions to demonstrate the flow because of a low clinical suspicion of a venous malformation. Even if a venogram was performed, it might not demonstrate any significant flow in these nondistensible OVMs. Orbital venous flow malformations were classified as superficial if they involved the periorbital skin, conjunctiva, or eyelid without outflow extension posterior to the equator of the globe. Deep orbital lesions were those located posterior to the equator of the globe with extraorbital involvement. Lesions with both superficial and deep orbital components were described as combined lesions.3

Seventeen patients required intervention. The patients were treated by surgical excision, carbon dioxide (CO2) laser ablation, embolization with platinum Guglielmi detachable coils (GDCs) (Target Therapeutics, Fremont, Calif), and percutaneous alcohol sclerotherapy depending on the distensibility and location of the lesion. Carbon dioxide laser ablation was performed transcutaneously in superficial lesions involving the skin and after surgical exposure in deeper lesions as a primary treatment without excision. A CO2 laser (model XL-40; Coherent Inc, Palo Alto, Calif) was used with a power setting of 2 W in the continuous mode.

Guglielmi detachable coil embolization was performed after intraoperative exposure of an orbital vein. An intraoperative venogram was obtained prior to the procedure. Following the access to the malformation through the exposed vein with a microcatheter, platinum GDCs were deposited.

Percutaneous sclerotherapy was performed after the puncture of the OVM under fluoroscopy and 3 mL of a combination 0.75:0.25 mixture of absolute alcohol and ethiodized oil (Ethiodol; Savage Laboratories, Melville, NY) was injected into the lesion without any occlusion using a 20-gauge angiocatheter.

Histopathological characteristics were examined in 11 patients. Main outcome measures were self-assessment of the patients regarding the improvement in symptoms or cosmesis as well as the objective improvement in visual acuity, extraocular muscle motility, and Hertel measurements. The patients were examined 1 day, 1 week, and 1 month postoperatively by the same physician (M.B.) and then followed up every 6 months. Six patients were lost to follow-up.

The cases of 18 of 22 patients (13 women and 9 men) reviewed in this study showed clinical or radiological evidence of distensibility. The remaining 4 patients had nondistensible OVMs. The age at onset of the initial signs and symptoms ranged from 5 months to 79 years, with a mean age of 32.4 years. The mean age at the initial manifestation of patients with distensible and nondistensible OVMs was 28.3 and 50.7 years, respectively. Eight patients (36.3%) had deep orbital lesions, 6 patients (27.3%) had superficial orbital lesions, and 8 patients (36.3%) had combined orbital lesions. All 3 patients with deep nondistensible orbital lesions were initially seen with a sudden onset of proptosis and pain or diplopia secondary to thrombosis or hemorrhage. Of the 18 patients with distensible lesions, no patients were initially seen with acute symptoms. Five of the 18 patients with distensible OVMs had deep lesions and 4 of them were initially seen with proptosis or pain increasing with the head in a dependent position. One patient with a deep distensible lesion was initially seen with diplopia and enophthalmos. Of the remaining 13 patients with superficial and combined distensible OVMs, 7 were initially seen with swelling at the location of the lesion.

Location of the lesion within the orbit was variable, but superior involvement was the most common (59%). Three patients had associated extraorbital venous malformations, involving the same side of the brain, face, and body as the orbital lesion.

Computed tomography (CT) was performed in 12 patients and typically showed tubular or lobulated homogeneous-nonhomogeneous-enhancing mass lesions. Four patients underwent magnetic resonance imaging. Magnetic resonance imaging showed enhancing mass lesions with variable signal intensities depending on the flow, presence of hemorrhage, or thrombosis within the lesion. Diagnostic orbital venogram was performed in 11 patients to confirm the diagnosis and to determine the routes of inflow and outflow as well as the extent of the lesion. Venogram was not performed if the lesion was small and superficial, if the OVM was not suspected clinically before the surgery, or if the patient was unable to afford the additional expense. Transfemoral cerebral angiography was performed in 7 patients with clinical signs or symptoms suggestive of an arterial flow lesion (ie, pulsation, bruit, and others) to rule out arteriovenous malformations.

Indications for intervention were cosmesis (7 patients), pain (6 patients), diplopia (2 patients), and optic nerve compression (2 patients). Five patients who did not receive any surgical intervention were observed. Seventeen patients required intervention. Four patients with

Table 1. Orbital Vascular Malformations

<table>
<thead>
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<th>No flow malformations</th>
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<tr>
<td>Lymphangiomas</td>
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<tr>
<td>Venous flow malformations</td>
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<tr>
<td>Distensible</td>
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<tr>
<td>Nondistensible</td>
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<tr>
<td>Mixed forms with venous and no flow components</td>
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<tr>
<td>Arterial flow malformations</td>
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<td>Arteriovenous malformations</td>
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nondistensible lesions were treated by surgical excision. Of the 13 patients with distensible lesions, 3 had superficial lesions that were treated by surgical excision (1 patient), CO₂ laser ablation (1 patient), or percutaneous alcohol sclerotherapy (1 patient) (Figure 1). Of the 7 patients with combined distensible lesions; 3 were treated by CO₂ laser ablation (Figure 2) and 2 by surgical excision. Two patients with combined distensible lesions (patients 16 and 17) had associated extraorbital venous malformations and they required multiple interventions including embolization of the venous malformations with platinum GDCs after surgical exposure.

Figure 1. Case 6. Distensible venous malformation of the left inferomedial orbit is shown before (A) and after (B) Valsalva maneuver. C, Computed tomographic scan of orbits and coronal section demonstrating oval enhancing lesion in the left inferomedial orbit. D, Venous phase of the cerebral angiogram, lateral projection, demonstrating the venous malformation (double arrowheads). E, Percutaneous puncture of the malformation under fluoroscopy and sclerotherapy, the alcohol-contrast mixture is injected into the lesion as noted in this lateral projection (double arrowheads). F, Complete resolution of the lesion 1-month postoperatively.

Figure 2. Case 11. A, Exposure of the lesion intraoperatively. B, Shrinkage of the lesion after carbon dioxide laser ablation.
Figure 3. Case 17. A, Computed tomographic view of the orbits, axial section, 6 years after subtotal surgical excision, showing an enhancing mass in the superior aspect of the right orbit (arrow) and additional abnormal enhancement in the right temporal area intracranially (double arrowheads). B, Orbital venogram in Waters projection, 19 years postoperatively, showing the intraorbital venous malformation (arrow). There is also reflux in multiple dilated irregular cutaneous and subcutaneous facial veins on the same side (double arrowheads). C, At the same session, Guglielmi detachable coil (GDC) embolization of the intraorbital venous malformation after intraoperative exposure of an orbital vein, access to the malformation through the exposed vein with a microcatheter (white arrow), and deposition of GDCs (black arrow). D, Platinum GDCs within the right orbit with no opacification of the intraorbital venous malformation (open arrow) 2 years after the embolization. A smaller subcutaneous venous malformation located over the bridge of the nose (black arrow) and another venous malformation over the right maxilla (double arrows) is also noted.

(Figure 3) for the deep component and CO₂ laser ablation or percutaneous alcohol sclerotherapy for the superficial portion. Three patients with deep distensible lesions had lateral orbitotomy and were treated with CO₂ laser ablation (1 patient), subtotal excision (1 patient), or ligation and cauterization (1 patient) of the vascular malformation.

Histopathological characteristics were examined in 11 patients. Two patients had associated intravascular papillary endothelial hyperplasia arising in a thrombosed vein.

Clinical and diagnostic features, treatment, and outcome of the patients are summarized in Table 2. Fourteen of the 17 patients treated had follow-up. Mean follow-up was 57.8 months (range, 2 months to 22 years). Eight patients had complete resolution of the signs and symptoms; the remaining 6 patients had marked improvement following surgery. Four of 11 treated patients who had distensible OVMs had long-term follow-up (>2 years). Postoperative ptosis developed in 2 patients. Transient skin blistering was noted at the site of injection in 1 patient after percutaneous alcohol sclerotherapy and resolved spon-
taneously in 1 month. No patient lost vision during follow-up. Best-corrected visual acuity was improved in 1 patient. Of the 5 patients who received no intervention and were only observed, 3 patients were lost to follow-up. The condition of the remaining 2 patients is stable, at a mean follow-up of 25.5 years.

**COMMENT**

The term “venous flow malformations” is applied to weakened segments of the orbital venous system of variable complexity including distensible lesions that have direct and rich communication with the venous circulation and nondistensible anomalies that have minimal communication with the venous system. The nondistensible vascular malformations seem to have a definite communication with the systemic venous circulation but to a much lesser extent than the distensible variety. To our knowledge, no large series of patients treated for OVMs, other than a series of 15 patients described by Lacey et al has been published. We report the clinical, radiological, and histopathological features, treatment, and outcome of 22 OVMs with the aim of describing a systematic approach for treatment.

Distensible and nondistensible OVMs showed different clinical features besides their different clinical and radiographic findings owing to distensibility. In our study mean age at manifestation was 28.3 years in patients with distensible OVMs. Nondistensible lesions were seen in an older-age group, with an average age of 50.7 years. Spontaneous orbital hemorrhage and thrombosis can be noted in all types of vascular malformations, but it is uncommon in distensible OVMs compared with the nondistensible variety. We noted thrombosis or orbital hemorrhage in 17.6% in the distensible and 100% in the nondistensible lesions. All except 1 patient with deep distensible OVMs were seen with proptosis or pain increasing with the head in a dependent position. One patient with a deep distensible lesion was seen with diplopia and was noted to have en-
ophthalmos in the sitting or standing position. Distension of the lesion with the head in a dependent position caused an enlargement of the orbit probably secondary to remodeling of the orbital bones as demonstrated by the CT scan (Figure 4). Most of the patients with superficial and combined lesions initially had swelling at the location of the lesion.

Histopathological examination was performed in 11 patients and this revealed dilated irregular veins lined by a single layer of endothelium with or without the presence of thrombus (Figure 5). Histopathological features of distensible and nondistensible lesions did not differ, although there was a higher incidence of thrombosis in the nondistensible ones. Histopathological examination of 2 nondistensible lesions with thrombosis showed associated intravascular benign endothelial hyperplasia, which is an unusual condition characterized by a benign proliferation of vascular endothelial cells that form papillary projections in the lumen of the blood vessel (Figure 6). It is believed to be a reactive response that develops secondary to a thrombus in vascular malformations or hemangiomas.6

Orbital CT with contrast enhancement is a reliable method of demonstrating OVMs.7 In our series orbital CT with contrast enhancement was the most frequently used radiological study to demonstrate the venous malformations. Orbital venography can be used to demonstrate their communication with the native venous circulation and routes of inflow and outflow as well as the extent of the lesions.5

The treatment of OVMs is largely conservative although extreme orbital pressure or intractable pain, profound orbital hemorrhage leading to visual deterioration, and cosmetic disfigurement can be indications for intervention.8 In our study all patients with nondistensible lesions and 72.2% of the patients with distensible lesions required intervention. The high percentage of cases requiring intervention may reflect the tertiary referral nature of our center.

To date there has been no definite standardization of treatment of OVMs. Thrombosed OVMs can be successfully treated by surgery if intervention is indicated.9 In our series 3 patients with deep thrombosed nondistensible OVMs required surgical treatment for optic nerve compression or severe pain and were treated by evacuation of clotted blood and subtotal excision of the lesion with good results. Recently, Gigantelli et al10 described an endoscopic transtheoidal decompression of a thrombosed OVM and evacuation of thrombus as a less invasive approach.

Table 2. Clinical, Radiological, and Histopathological Features of Orbital Venous Flow Malformation, Treatment, and Outcome (cont)

<table>
<thead>
<tr>
<th>Distensibility Depth</th>
<th>Patient No./ Age, y</th>
<th>P sx</th>
<th>Location</th>
<th>Venogram</th>
<th>Ix</th>
<th>Treatment</th>
<th>Pathological Feature</th>
<th>Follow-up</th>
<th>Outcome</th>
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<td>Distensible Combined 15/40 Facial vascular lesions Diffuse Prominent vein in orbit and middle cranial fossa on the same side Cosmesis CO2 laser ablation NA NA NA</td>
<td>16/28</td>
<td>Swelling, diplopia Superior</td>
<td>Large sinusoidal venous lesion draining into angular and facial vein, cavernous sinus, and petrosal sinus Pain Lat and anterior orbitotomy—excision with CO2 laser Surgical exposure and GDC embol CO2 laser ablation (upper eyelid) CO2 laser ablation (upper eyelid) Congested dilated thin-walled vessels 11 y* Improvement Ptosis and diplopia</td>
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<td>Deep 18/20</td>
<td>Proptosis Superior and intracranial Inferior orbital vein not opacified Pain Lat orbitotomy—ligation and cauteterization of lesions</td>
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<td>19/25</td>
<td>Pain Superior and intracranial NA Pain Lat orbitotomy—subtotal excision Surgical exposure and GDC embol Thrombosed varix with early BPEH Organizing blood clot 18 y Resolution</td>
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<td>20/39</td>
<td>Pain Superior and intracranial NA Pain Lat orbitotomy—CO2 laser ablation</td>
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<td>21/61</td>
<td>Proptosis Superior and intracranial Varicoce enlargment of intracranial SOV NA Observation NA 23 y Stable</td>
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<td>22/37</td>
<td>Diplopia Superior apex Sinusoidal venous lesion filled via SOV NA Observation NA NA NA</td>
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Abbreviations: BPEH, benign papillary endothelial hyperplasia; Co2, carbon dioxide; embol, embolization; GDC, Guglielmi detachable coil; Ix, indication; lat, lateral; NA, not applicable; ON, optic nerve; Psx, presenting symptom; SOV, superior ophthalmic vein.

*Nine years’ follow-up after the last intervention for the orbital venous flow malformation, 2 months’ follow-up after the last CO2 ablation of the eyelid lesion.
†Two years’ follow-up after the last intervention for the orbital venous flow malformation.
Superficial and combined lesions can be treated by CO₂ laser ablation either transcutaneously or after surgical exposure of the lesion without surgical excision. It is particularly useful if the lesion is extensive and diffuse. Carbon dioxide laser ablation has been used as an adjunct to surgical excision in the past, but we believe that it can also be successfully used as a primary treatment in selected superficial and combined OVMs as a less invasive approach. If the lesion is small and circumscribed, it can be excised surgically with the help of the CO₂ laser. The CO₂ laser can also be helpful for treatment of deep OVMs. One patient with an intraconal distensible OVM underwent lateral orbitotomy and CO₂ laser ablation of the lesion with a successful result.

Although percutaneous alcohol sclerotherapy of venous malformations in various anatomical sites were found to be effective and safe, experience with OVM has been limited and this might be explained by the concern about the risk of optic nerve injury secondary to leakage of alcohol within the orbit. We describe a 10-month-old patient with a superficial distensible OVM treated by percutaneous alcohol sclerotherapy. Following treatment, this patient had self-limiting skin blistering over the lesion and the OVM completely resolved in 3 months. It is our opinion that percutaneous alcohol sclerotherapy can be applied to superficial OVMs with caution as a less invasive treatment alternative. Best results have been reported with small well-defined OVMs instead of large infiltrating lesions in other parts of the body.

Deep distensible OVMs have been traditionally treated by surgical excision. Surgical excision can be facilitated by the use of clips or by the recently described technique of embolization with cyanoacrylate glue after surgical exposure to prevent bleeding during excision. Embolization with cyanoacrylate glue is not free of complications including vision loss. The lesion with the material must be removed surgically since it causes a foreign body reaction if it is left in the orbit. Hard consistency of the glue can also lead to difficulty in surgical dissection of the embolized lesion. Nonadhesive liquid agents such as ethylene vinyl alcohol copolymer (Onyx; Microtherapeutics Inc, Irvine, Calif) have been recently shown to be useful in the treatment of arteriovenous malformations. These can be potentially useful for future treatment of OVMs.

Endovascular treatment by platinum GDC embolization is a less invasive alternative to surgical excision for deep distensible OVMs. Preoperative or intraoperative venography can identify the patients for whom embolization is appropriate. Takechi et al reported a case of an orbital varix treated with percutaneous transfemoral venous catheterization and embolization with platinum GDCs. This approach can be technically difficult owing to developmental variations. Embolization of OVMs after surgical exposure has the advantage of avoiding difficult catheterization and limiting traumatic dissection. Weill et al reported a case of an orbital varix treated by embolization with platinum GDCs after surgical exposure with good result. In our series 2 patients with complex OVMs were treated by embolization with platinum GDCs after surgical exposure. One of these patients (case 17) who had a history of subtotal excision of the lesion during childhood underwent 2 endovascular treatments with embolization. The patient’s symptoms improved markedly after the second emboliza-
tion. The follow-up venogram showed complete occlusion of the OVM 2 years postoperatively (Figure 3D).

The other patient (case 16) had endovascular treatment for the deep orbital component of the lesion and also had multiple ablations with CO₂ laser for the superficial portion, with considerable improvement of the signs and symptoms. Embolization is more useful for saccular or segmental venous dilations than for tangled plexus of venous channels. Embolization with platinum GDCs is not a preferred choice of treatment for the superficial lesions since it shows its effect by inducing thrombosis in the lesion without an actual decrease in the size of the lesion, and this may not be cosmetically acceptable.

Our preferred treatment choices can be summarized by the following criteria:

1. For distensible or nondistensible OVM without intractable pain, orbital hemorrhage leading to visual deterioration, or cosmetic disfigurement: observation
2. For distensible OVM:
   a. Superficial well-circumscribed
      i. Percutaneous alcohol sclerotherapy
   b. Superficial diffuse: CO₂ laser ablation; transcrotaneous or after surgical exposure
   c. Deep
      i. Embolization with platinum GDCs
      ii. Surgical exposure followed by CO₂ laser ablation
      iii. Surgical excision facilitated by the use of clips or prior embolization with cyanoacrylate tissue glue
3. For nondistensible OVM: Lesions associated with thrombosis and/or hemorrhage:
   a. Without intractable pain, threatened optic nerve compression, or severe proptosis: Observation if the diagnosis is made clinically
   b. With intractable pain, optic nerve compromise, severe proptosis, or repeated/nonresolving episodes of thrombosis: Surgical exposure and evacuation of clot with subtotal excision

CONCLUSIONS

If intervention is indicated, the aim should be to consider the least invasive method as a first-line treatment since these lesions are benign and symptomatic treatment is satisfactory in most patients. Over the years, these malformations have been treated by surgical excision facilitated by different methods. We believe the patients who require intervention should be given the choice of a less invasive method such as CO₂ laser ablation and percutaneous alcohol sclerotherapy for superficial and combined lesions and endovascular treatment by platinum GDC embolization for deep lesions. In cases of acute thrombosis or hemorrhage associated with severe pain, proptosis, or visual loss, surgery may be required to remove the thrombosed lesion or hematoma. The complex venous lesions fall into a clinical spectrum of vascular malformations affecting the orbit, face, and brain; they are difficult to treat and treatment must be customized for each patient individually.

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