

# Venous Malformation of the Orbit

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Terminologic and clinical confusion of vascular anomalies persists in the literature. Vascular anomalies may occur anywhere in the body, and treatment becomes complicated when they occur in the craniofacial region. We present here the case of a patient with an orbital venous malformation causing ocular dystopia, to illustrate the importance of proper terminology and accurate diagnosis through appropriate imaging investigations. We show that a satisfactory outcome can be achieved using standardized craniofacial surgical approaches through hidden incisions.

## Report of a Case

A 17-year-old female patient previously underwent excision of a left orbital lesion through a temporal skin incision at another institution. The procedure was abandoned when profuse bleeding was encountered. The histology of the tissue obtained was reported as a "cavernous hemangioma" (sic). The patient was subsequently referred to the Centre for the Study & Treatment of Vascular Birthmarks for further management.

Physical examination revealed ocular dystopia, with the left globe being 5 mm higher than the right as well as an external scar in the temple (Fig 1). There was normal visual acuity and extraocular movement without diplopia. Review of the original computed tomography (CT) scan showed a 2-cm lesion lying on the orbital floor displacing the globe superiorly (Fig 2A). The lesion was closely applied to a bony

spur arising from the inferior orbital margin, although there was no radiologic evidence of osseous infiltration (Fig 2B). Magnetic resonance imaging (MRI) showed that the orbital mass was intimately related to the inferior rectus muscle and displacing it slightly medially, although there was a clear plane between these structures. The lesion was hyperintense on T2-weighted images (Fig 3), and it showed marked gadolinium contrast enhancement on T1-weighted sequences. These CT and MRI features were consistent with a venous malformation.<sup>1,2</sup> A superselective carotid angiogram showed normal arterial anatomy and blood flow.

The patient underwent resection of the venous malformation through a subciliary incision. Intraoperatively, scar tissue was noted around the lateral wall of the orbit. A subperiosteal dissection was carried out along the lateral and inferior orbital margins, over the floor and the lateral wall of the orbit. There was bony hypertrophy of the floor and the inferior orbital margin, resulting in reduction of the orbital volume and the bony lip, respectively. Excision of the lesion was facilitated by removal of the bony lip at the inferior orbital margin. The lesion was removed easily without injury to the inferior rectus and inferior oblique muscles or the intraconal contents. Removal of the excess bone in the floor of the orbit was performed to normalize the orbital volume, allowing the globe to return to a satisfactory position.

The postoperative course was uneventful with no complications such as disturbance of the visual acuity or diplopia. Histologic examination of the excised specimen showed features consistent with venous malformation.<sup>3-6</sup> There were large thin-walled vessels scattered within a fibrous stroma. The channels were lined by flat endothelium and contained smooth muscle within their walls (Fig 4) revealed by immunoperoxidase staining for smooth muscle actin. In addition, there were foci of capillary-sized vessels similar to those observed in the original biopsy. Within the specimen, there also were small bony trabeculae and focal hemosiderin deposition (indicative of hemorrhage) probably due to previous surgery or thrombosis.

At follow-up 4 years after surgery, there was no clinical or MRI evidence of recurrence of the venous malformation. The left globe remained in a satisfactory position with no visual dysfunction (Fig 5). The temporal scar had improved, although it remained conspicuous.

## Discussion

Terminologic and clinical confusion of vascular anomalies has been responsible for improper diagnosis, illogical treatment, and misdirected research efforts.<sup>3,7</sup> Traditionally, the term *hemangioma* and *angioma* have been indiscriminantly used to describe nonproliferative, developmental vascular malformations as well as vascular lesions that expand via true

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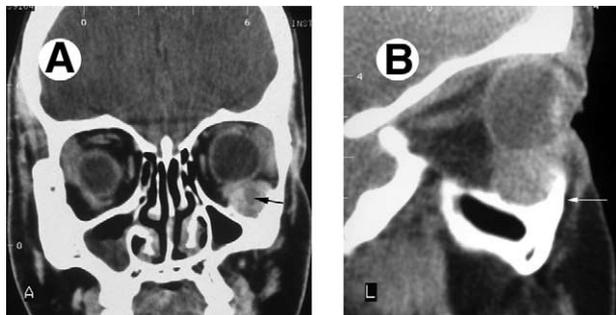


**FIGURE 1.** A 17-year-old female patient presented with ocular dystopia with the left globe being 5 mm higher than the right. Note the conspicuous temporal scar resulting from previous attempted excision performed elsewhere.

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cellular hyperplasia. Commonly applied morphologic modifiers such as “cavernous,” “racemose,” “simplex,” “capillary,” and “capillary-cavernous,” many dating from the time of Virchow in the 1800s, often add more confusion than enlightenment.<sup>5</sup>

In the literature, almost every type of vascular anomaly has, at some point, been called hemangioma.<sup>8</sup> The term *hemangioma* should be restricted to a rapidly growing vascular tumor of infancy,<sup>4,6,8</sup> com-



**FIGURE 2.** (A) An axial computed tomography scan showing a 2-cm mass lesion (arrow) closely applied to the hypertrophic left orbital floor displacing the globe superiorly. (B) Sagittal computed tomography scan showing the lesion was situated above the orbital floor and against the bony lip arising from the hypertrophic inferior orbital margin (arrow).

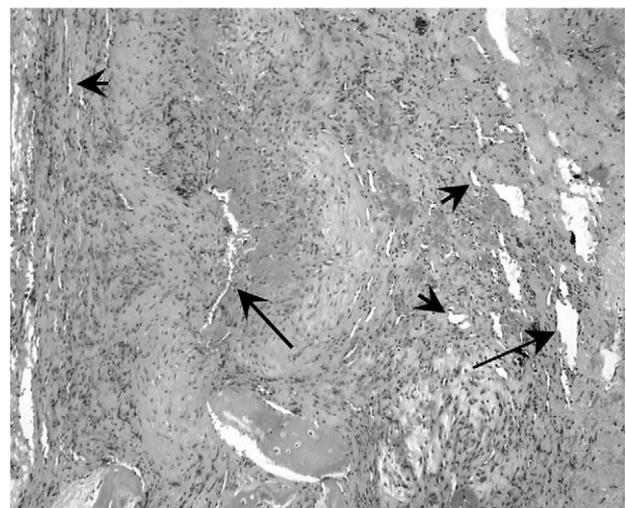
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**FIGURE 3.** The lesion (arrow) was hyperintense on T2-weighted magnetic resonance image.

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monly known as *strawberry nevus*. This benign tumor is usually not present at birth, appears during the first 2 weeks of life, and undergoes spontaneous regression within 5 to 12 years.<sup>4,7,8</sup> Hemangiomas do not occur during adolescence or adulthood.<sup>4,8</sup> The other major category of vascular anomalies are vascular malformations, which are composed of dysplastic vessels lined by quiescent endothelium.<sup>3-6</sup> Vascular malformations are present, although they are not always detected, at birth. These lesions grow propor-



**FIGURE 4.** Photomicrograph showing large thin-walled vessels scattered within a fibrous stroma. The channels are lined by flat endothelium and contain smooth muscle within their walls (long arrows). There are also foci of capillary-sized vessels (short arrows) consistent with organizing thrombosed vessels that are frequently observed in venous malformation (hematoxylin and eosin stain, original magnification,  $\times 20$ ).

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**FIGURE 5.** At follow-up 3.5 years after surgery, there was no clinical or magnetic resonance imaging evidence of recurrence of the venous malformation. The left globe was in a satisfactory position with no visual dysfunction.

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tionately with the patient, and sometimes expand in response to hormonal changes (eg, puberty and pregnancy), trauma, or incomplete excision, but they never regress spontaneously.<sup>4,7</sup>

For 2 decades, hemangioma and vascular malformations have been distinguished as different biologic entities by clinicohistopathologic,<sup>3,5,6,9-11</sup> *in vitro*,<sup>12</sup> and radiologic<sup>1,2</sup> studies. In addition, a number of cellular and extracellular markers have been used to differentiate between hemangioma and vascular malformations.<sup>9-11</sup>

The biologic classification of vascular anomalies into hemangioma and vascular malformations, first proposed by Mulliken and Glowacki<sup>3</sup> in 1982, has now been adapted by the International Society for the Study of Vascular Anomalies (Berlin, 1998). This classification categorizes vascular anomalies into vascular tumors and vascular malformations. Hemangioma, the most common tumor of infancy, has been well described,<sup>8</sup> and its biology was characterized more recently.<sup>4,6,8-11,13-17</sup> Other less common vascular tumors include kaposiform hemangioendothelioma<sup>6,8,18,19</sup> and congenital tufted angioma.<sup>6,8,19</sup> Vascular malformations are true developmental anomalies that can be further subcategorized according to the channel mor-

phology with the capillary, vein, artery, or lymphatic vessel being involved either singly (simple malformations) or in combination (combined malformations).<sup>8</sup> Vascular malformations are also classified according to rheology as either slow-flow or fast-flow.<sup>3,4,8</sup> Clinical features of a high-flow lesion include a blush, thrill, and bruit. A pulsatile globe may be associated with an intraorbital lesion, as well as bony destruction or deficiency.<sup>8</sup> In contrast, a low-flow lesion may be associated with pressure symptoms, pain secondary to thrombosis, and bony or soft tissue over-growth.<sup>8</sup>

The patient presented here highlights the importance of precise terminology, accurate diagnosis, and appropriate investigations, all critical for proper treatment. The diagnosis of a vascular anomaly can be made clinically on the basis of careful history and physical examination in a majority of cases without resorting to invasive studies.<sup>20</sup> However, when the lesions occur in the craniofacial region, appropriate investigations may be necessary. Various imaging techniques, including ultrasonography, technetium-labeled red blood cell scintigraphy, angiography, CT, and MRI, have been used to study vascular anomalies.<sup>1,2,21-24</sup> However, MRI with gadolinium enhancement is considered the single most informative investigation. It verifies the clinical diagnosis, shows the flow characteristics, and shows accurate anatomic details of the lesion.<sup>22</sup> CT scanning provides information regarding bony involvement or associated bony anomaly. An angiogram is essential for high-flow lesions. With this imaging available, a biopsy is rarely required.

The histologic features described in this report are those of venous malformation. The presence of bony trabeculae and focal hemosiderin deposition within the lesion may be caused by previous trauma/surgery or thrombosis. These features are often observed in a low-flow lesion such as venous malformation. The presence of groups of capillary-sized vessels in venous malformation should not be confused with those seen in a hemangioma. These tiny vessels represent recanalization in thrombosed vessels that commonly occur in these lesions.<sup>6</sup>

Using standardized craniofacial approaches, lesions in and around the orbit can be removed through hidden incisions with or without appropriate orbitotomies.<sup>25</sup> Those in the orbital floor, such as the one presented in this report, can be excised through a subciliary or conjunctival incision,<sup>26</sup> and if necessary, an inferior marginal orbitotomy,<sup>25</sup> to improve access. In addition, a lateral marginal orbitotomy<sup>25</sup> with removal of the anterior part of the lateral orbital wall may be required to control troublesome bleeding during excision and to remove a larger lesion, especially

if it encroaches on the lateral orbital wall, in which case a coronal incision is required.

Low-flow vascular anomalies such as venous malformation may be associated with osseous hypertrophy or may even be entirely intraosseous.<sup>7</sup> Although it is generally believed that hypertrophy of the bone adjacent to a venous malformation results from an increased local blood flow, it may be related to the mild pressure phenomenon stimulating the growing periosteum. This phenomenon is seen in other inflammatory conditions where increased layering of the paracortical bone is observed in the younger patient. In the case reported here, both the mass effect of the lesion and the reduction of the orbital volume secondary to bony hypertrophy contributed to the ocular dystopia. It is important to remove the excess bone to normalize the orbital volume to achieve a satisfactory correction of the ocular dystopia.

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