Spontaneous Hemorrhage From Orbital Cavernous Hemangioma Resulting in Sudden Onset of Ophthalmopathy in an Adult

—Case Report—

Junkoh YAMAMOTO,1 Mayu TAKAHASHI,1 Yoshiteru NAKANO,1 Takeshi SAI TO,1 Takehiro KITAGAWA,1 Kunihiro UETA,1 Ryo MIYAOKA,1 and Shigeru NISHIZAWA1

1Department of Neurosurgery, University of Occupational and Environmental Health, Kitakyushu, Fukuoka

Abstract

A 61-year-old woman presented with a very rare case of spontaneous bleeding from an orbital cavernous hemangioma manifesting as sudden onset of ophthalmic pain, proptosis, diplopia, and nausea. Magnetic resonance (MR) imaging and computed tomography (CT) revealed an intraconal, well-demarcated cystic mass with hemorrhage. The mass was immediately removed through a transcranial approach. Histological examination showed that the mass was a cavernous hemangioma. Ophthalmopathy was resolved by surgery. Orbital cavernous hemangioma is very common in adults, but spontaneous bleeding is extremely rare relative to intracranial cavernous hemangiomas. This rare case emphasizes the importance of careful neuroimaging evaluation with MR imaging and CT for diagnosing orbital cavernous hemangioma associated with spontaneous bleeding.

Key words: orbital tumor, intratumoral hemorrhage, proptosis, diplopia, vascular malformation

Introduction

Cavernous hemangiomas are one of the most common primary orbital mass tumors in adults.7) Orbital cavernous hemangiomas may be incidentally detected by computed tomography (CT) or magnetic resonance (MR) imaging performed for unrelated conditions, which usually show either no growth or slow growth. Thus, orbital cavernous hemangiomas, which are considered benign, differ from other orbital vascular lesions such as lymphangiomas and hemangiopericytomas.10,12) No malignant transformation of orbital hemangiomas has been reported. Therefore, the subset of patients with asymptomatic orbital masses that are clinically and radiologically compatible with cavernous hemangiomas may never require surgery.8,11)

Here, we report the case of an adult patient with orbital cavernous hemangioma who suffered sudden onset of ophthalmopathy due to spontaneous bleeding from an orbital cavernous hemangioma. The symptoms of the patient were relieved by surgery.

Case Report

A 61-year-old otherwise healthy woman suffered sudden onset of ophthalmic pain with proptosis and diplopia of the left eye as well as nausea and vomiting. The patient visited the emergency department of a local hospital. Emergency CT detected an intraorbital tumor. Two days later, she was referred to our university hospital for further examination. On admission, she complained of left ophthalmic pain, diplopia, and nausea. Physical examination showed proptosis (22 mm) with lid swelling and a partial visual field defect in the left eye (Fig. 1A). Ocular movements of the left eye were also slightly limited in all directions. Axial CT revealed an extensive mass with a large high density area located in the left orbit, and coronal CT showed an intraconal mass occupying the lower temporal part of the left orbit; the mass appeared to push the optic nerve superonasally (Fig. 1B, C). No phlebolith within the mass and no bony spur arising from the inferior orbital margin was identified (Fig. 1C, D). MR imaging also revealed a well-demarcated intracranal mass with partial low signal intensity within the mass as well as a low signal intensity capsule on T2-weighted imaging, and partial high signal intensity on T1-weighted imaging, with a fluid-fluid level on both sequences (Fig. 2A–C). These heterogeneous signal intensities indicated acute to subacute hemorrhage within the mass. MR imaging with contrast medium failed to show enhancement of any major part of the mass (Fig. 2D).

Under a preoperative diagnosis of hemorrhagic neurinoma and cavernous hemangioma, surgery was per-
formed via a transcranial approach (frontotemporal craniotomy) 9 days after the symptom onset. The mass appeared dark purple, and the extended cystic portion was surrounded by a thin wall (Fig. 3). The mass was shrunken after an “old” collection of blood within it was removed. The thin wall of the mass was partially adhered to the surrounding tissues, including muscular structures and nerves, so subtotal resection of the wall was performed to prevent postoperative complications. Elastic and fibrous portions of the mass could not be confirmed within the surgical fields. Postoperative MR imaging revealed that the left orbital mass had been removed and left exophthalmos was resolved (Fig. 4). Histological examination showed thin-walled, blood-filled dilated vascular spaces lined by flattened endothelial cells (Fig. 5A, B). These dilated vascular channels also showed mild inflammatory infiltration and mild overgrowth of fibroblasts in the adjacent adipose tissue without thickening of the circumferential blood vessels (Fig. 5A–D). Some inflammatory infiltrated cells contained hemosiderin (Fig. 5E). The histological diagnosis was cavernous hemangioma. After surgery, the patient was relieved of ophthalmic pain, proptosis, and nausea; the visual field defect was also corrected. However, diplopia improved only gradually.

**Discussion**

The growth of orbital cavernous hemangiomas is extremely slow, and some cases may not grow at all after a certain age.\(^9\) The most common presenting sign and symptom is painless proptosis.\(^1\) However, this symptom has a slow onset and often remains unnoticed by the patient.\(^1,2,7,10\) In contrast to the frequent hemorrhaging observed with intracranial cavernous hemangiomas, spontaneous intraorbital hemorrhage from orbital cavernous hemangiomas is extremely rare. Only 3 previous cases of spontaneous intraorbital hemorrhage caused by an orbital cavernous hemangioma have been reported. All these cases were evaluated preoperatively using CT.\(^19\) The present case of spontaneous intraorbital hemorrhage caused by an orbital cavernous hemangioma was detected with MR imaging.

Generally, vascular malformations of the orbit are classified on the basis of hemodynamic concepts as follows. Type 1 (no flow) lesions have essentially little connection to the vascular system and include lymphangiomas or combined venous lymphatic malformations. Type 2 (venous flow) lesions present either as distensible lesions with direct and rich communication with the venous system or as non-distensible anomalies with minimal communication with the venous system. Type 2 includes venous malformations, varices, and orbital venous anomalies. Type 3 (arterial flow) lesions include arteriovenous malformations characterized by direct anti-grade high flow through the lesion to the venous side.\(^9\) Occasional findings of small feeding arteries and delayed contrast pooling using conventional, prolonged digital subtraction...
Fig. 3 Intraoperative photograph. Frontotemporal craniotomy exposed the cystic tumor (T) that had extended between the left superior levator palpebrae and superior rectus muscle (A) and the left lateral rectus muscle (B).

Fig. 4 Axial T₂-weighted (A) and coronal fat-saturated T₁-weighted with contrast medium magnetic resonance images (B) showing that the left orbital mass had been removed and exophthalmos was resolved. An irregular, enhanced structure was found inferior to the optic nerve at the orbital apex, but residual lesions or postoperative changes were difficult to determine (arrows).

angiography techniques have been reported. Orbital cavernous hemangiomas are thought to be of venous origin and are classified as Type 3 low-flow arteriovenous malformations.¹²,¹⁴,¹⁵

Orbital hemorrhage due to orbital venous anomalies is not infrequent.⁶,¹⁵ The clinical features of orbital venous anomalies are known to include worsening proptosis with increased venous pressure induced either by a Valsalva maneuver or by lowering of the head.¹⁵ CT and MR imaging may show enlargement of an orbital venous anomaly during a Valsalva maneuver or venous compression.⁶,¹⁴,¹⁵ Phleboliths and bony spurs arising from the orbital margin are also characteristic of orbital venous anomalies.¹³,¹⁵ Histologically, orbital venous anomalies consist of irregular, dysmorphic venous channels presenting with abnormally thickened vessel walls.⁶,⁹ A recent study reported that orbital hemorrhage due to an orbital venous anomaly resolved spontaneously in approximately 1–2 weeks, so conservative treatment and repeat CT are recommended for such cases.⁶

In our present study, there was no tendency for the ophthalmopathy to resolve during the 9 days preceding surgery. Our case presented with neither phleboliths nor abnormal bony changes on CT. Histological examination revealed lesions consisting of thin-walled, blood-filled vessels with partially fibrous stroma, all of which are in agreement with a cavernous hemangioma.⁵,⁹,¹⁰ The previous 3 reported cases of intraorbital hemorrhage caused by cavernous hemangiomas manifested as sudden onset of exophthalmos (either with pain or painless), ptosis, and diplopia.¹⁶ Surgery was performed in all 3 cases and cavernous hemangioma was histologically confirmed. In 2 of these 3 cases, surgery was performed 3 or 4 weeks after the symptom onset because clinical or radiological findings were unchanged. Spontaneous resolution may be difficult in orbital hemorrhage from cavernous hemangioma, unlike hemorrhage from orbital venous anomalies.
which have direct and rich communication with the venous system. If orbital hemorrhage caused by a cavernous hemangioma is suspected by neuroimaging examination, and there is no tendency toward spontaneous recovery, we recommend surgical treatment at an early stage.

The mechanism underlying growth or bleeding of orbital cavernous hemangiomas remains unclear. Histologically, cavernous hemangiomas are classically defined as large, dilated vascular channels containing blood and occasional thrombi, which are lined by flattened or attenuated endothelial cells and intervening fibrous septae within a pseudocapsule. Local hemodynamic disturbances and hypoxia, caused by low blood flow or ischemia, are considered to induce intrapapillary endothelial hyperplasia and capillary formation, thus initiating the slow enlargement of cavernous hemangiomas. The histopathological findings in our present case showed an expanding, large, and dilated vascular channel containing a fluid hematoma without a thick, fibrous interstitium. In addition, the capsule of the mass was attached to the surrounding tissues and showed partial inflammatory changes. Therefore, we suspect that the inflammatory event occurred in the small capillaries within the orbital cavernous hemangioma during the early stages of its growth. The inflammatory event disturbed and obstructed the venous circulation within the orbital cavernous hemangioma. Hemorrhage within the mass subsequently occurred, and the mass suddenly expanded because of the immature and fragile fibrous interstitium.

Cavernous hemangiomas are very common primary orbital masses in adults and are considered benign. Spontaneous bleeding of an orbital cavernous hemangioma is extremely rare. However, once bleeding occurs, severe ophthalmopathy may suddenly develop, necessitating emergency surgery, as observed in the present case. Therefore, precise diagnosis of orbital cavernous hemangiomas by careful observation using MR imaging is essential.

References


Address reprint requests to: Junkoh Yamamoto, MD, PhD, Department of Neurosurgery, University of Occupational and Environmental Health, 1-1 Iseigaoka, Yahatanishi-ku, Kitakyushu 807–8553, Japan.
E-mail: yama9218@med.uoeh-u.ac.jp

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