

# A Cavernous Venous Malformation of the Orbit Mimicking an Idiopathic Orbital Inflammation

## Abstract

Orbital cavernous venous malformations (CVMs) are usually slow progressing. Multiple CVMs, bilateral orbital CVMs, and acute presentations are rare. We present a rare, bilateral, orbital CVM with acute painful visual loss in the left eye. The initial clinical presentation mimicked an idiopathic orbital inflammation. Orbital magnetic resonance imaging revealed its rare location at the left orbital apex. Finally, pathology confirmed the presence of an intralesional hemorrhage of a CVM.

**Keywords:** *Cavernous venous malformations, idiopathic orbital inflammation, orbital*

## Introduction

Cavernous venous malformations (CVMs) of the orbit, formerly known as orbital cavernous hemangiomas, are the most common, primary, orbital lesions of adults.<sup>[1]</sup> CVMs occur more often in middle-aged women.<sup>[2]</sup> Typically, orbital CVMs are slow-growing and noninfiltrative lesions. The most common presenting complaints are retrobulbar or periorbital pain and visual loss,<sup>[2]</sup> while the most common presenting sign is proptosis. CVMs are usually located in the intraconal space.<sup>[2]</sup>

We report a rare case of a bilateral orbital CVM with an intralesional hemorrhage in the left eye mimicking an idiopathic orbital inflammation.

## Case Report

A 60-year-old woman presented with severe headache and visual loss in the left eye for 2 days. Initially having the headache at the left temporal region and around the left eye, she had developed the visual loss in the left eye after a few hours. The patient denied having pain on eye movement, fever, jaw claudication, or weight loss. She also reported no history of autoimmune diseases or recent trauma.

The patient went to her primary care hospital. A computed tomography (CT)

scan of the orbit revealed a left orbital-apex mass adhering to the left superior rectus and medial rectus muscles and a right orbital apex mass [Figure 1]. She was diagnosed as having an orbital pseudotumor and was referred to our hospital for further management. Her visual acuity in the right eye was 20/40 and counting fingers in the left eye. An eye examination found a relative afferent pupillary defect as well as a mild limitation of the lateral rectus movement in the left eye. There was no lid swelling or proptosis, and the fundus and disc appeared normal. We observed that the patient also had a blue–green soft nodule on her left cheek [Figure 2]. We reviewed her CT scan and noticed a homogeneous lesion on her left cheek [Figure 3]. Consequently, we thought the cheek lesion might be a hemangioma related to her orbital lesions. We requested magnetic resonance imaging (MRI) of the brain and orbit.

The MRI scans revealed an ill-defined intraconal mass at the medial portion of the left orbit causing a lateral pressure effect on the left optic nerve. The mass had an iso-to-low signal intensity (SI) on T1W, a low SI on T2W, and peripheral patchy enhancement. The mass involved part of the left superior oblique and left medial rectus muscles. However, part of the central portion showed a particularly low SI on T2W without enhancement, which was suspicious of a

**Niphon Chirapapaisan, Chanon Ngamsombat<sup>1</sup>, Jantima Tanboon<sup>2</sup>, Pornsuk Cheunsuchon<sup>2</sup>, Sugamon Koohasawad<sup>3</sup>**

*Departments of Ophthalmology, <sup>1</sup>Radiology and <sup>2</sup>Pathology, Faculty of Medicine, Siriraj Hospital, Mahidol University, <sup>3</sup>Department of Ophthalmology, Prasat Neurological Institute, Bangkok, Thailand*

**Address for correspondence:**  
Dr. Niphon Chirapapaisan,  
Department of Ophthalmology,  
Faculty of Medicine, Siriraj  
Hospital, Mahidol University,  
Siriraj, Bangkok Noi,  
Bangkok 10700, Thailand.  
E-mail: niphon.chi@mahidol.  
ac.th

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hemorrhagic component. Moreover, there was another small, lobulated, intraconal nodule at the right orbital apex that had a low SI on T1W and a high SI on T2W with homogeneous enhancement. It was located just lateral to the right optic nerve without intraocular muscle involvement [Figure 4].

A neurosurgeon was consulted for tissue diagnosis and management of the lesion in the left orbit. The patient underwent left craniotomy and orbitotomy with the aid of CT navigation. The finding was a pink, ill-defined, soft, sticky, and easily bleeding mass at the medial region of the left orbital apex. There were some old blood clots and a few large blood vessels supplying the mass. The mass adhered to the left medial rectus muscle, the left superior oblique muscle, and the optic nerve. Tumor removal was performed. The pathology showed collapsed, thin-walled, vascular channels associated with a dilated area containing organized thrombus, all of which were compatible with a hemangioma with an intralesional hemorrhage [Figure 5].

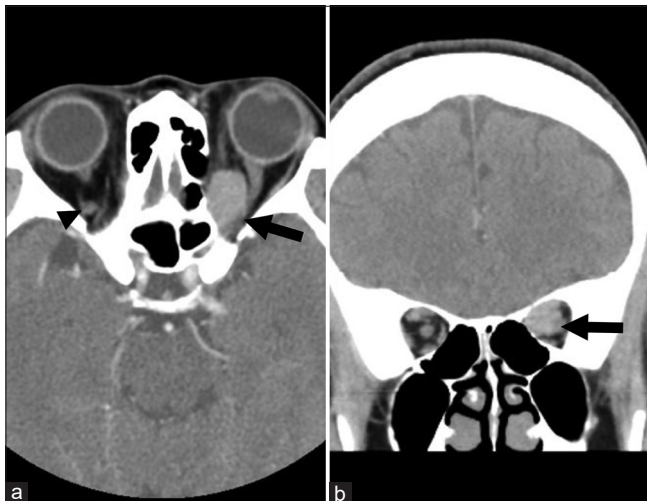


Figure 1: The computed tomography scan of the orbit showed a homogeneous, enhancing mass at the left orbital apex (Fig 1.a) adhering to the left superior rectus (Fig1.b) and medial rectus muscles (Fig1.a) and causing lateral compression on the left optic nerve (arrow). Furthermore, there was a small enhancing nodule at the right orbital apex (Fig 1.a, arrow head)

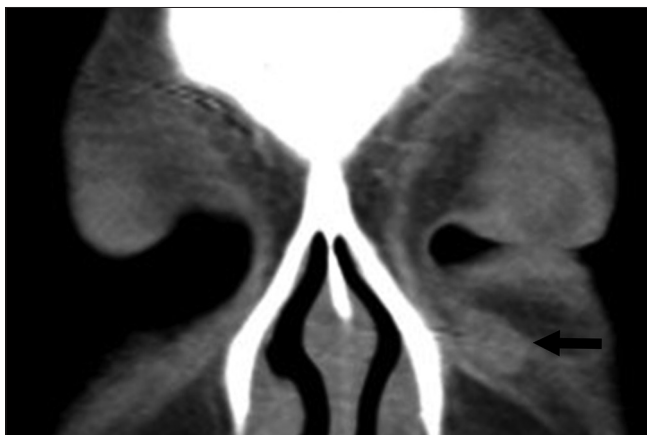


Figure 3: A homogeneous lesion at the infraorbital area of the left cheek was just lateral to the left nasal ala (arrow)

The patient was diagnosed as having a CVM with an intralesional hemorrhage at the left orbital apex. Two days after the surgery, her vision improved from counting fingers to 20/500 in the left eye. The best-corrected visual acuity of the left eye 20/50 was achieved 1 month postoperatively, and there was no pain nor any limitation of the patient's eye movement. She attended all follow-up sessions. Two years after the procedure, her visual acuity was 20/25 in both the eyes. An MRI scan at that time revealed no change in the CVM in the right eye and no residual tumor in the left eye.

## Discussion

We presented a patient with an acute visual loss and pain around the left eye. The clinical features and orbital imaging of this patient suggested an idiopathic orbital inflammation. However, we noticed identical that painless lesions were present at both the right orbit and the left cheek. This observation led us to consider that the lesions might instead be part of a multiple CVM. After reviewing the patient's CT scans, we confirmed diagnosis with an MRI scan and a pathology investigation.



Figure 2: A blue-green soft nodule was present on the left cheek

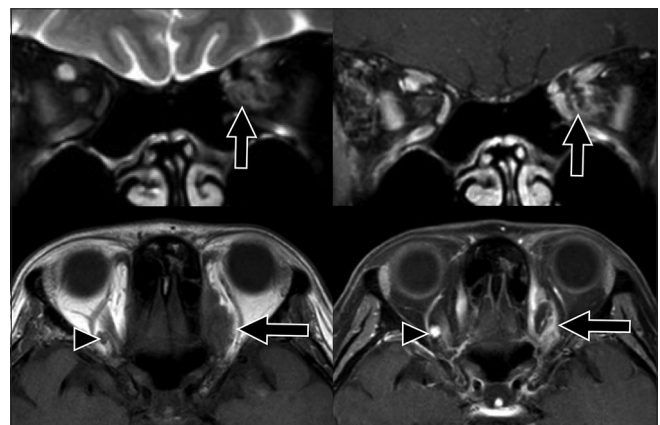
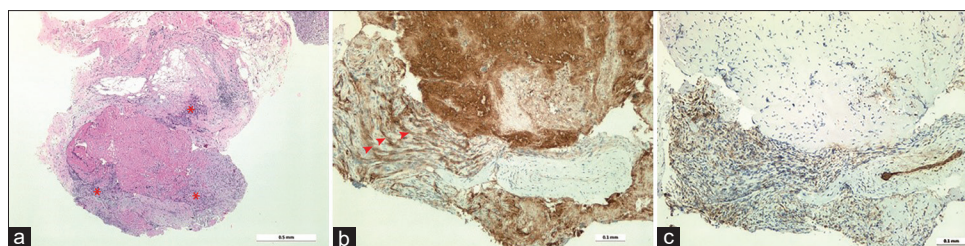


Figure 4: The magnetic resonance imaging scan revealed that an ill-defined, intraconal mass at the medial portion of the left orbit (arrow) was causing a lateral pressure effect on the optic nerve. The mass showed iso-to-low signal intensity on T1W, low signal intensity on T2W, and peripheral patchy enhancement. It involved part of the left superior oblique and left medial rectus muscles. However, some central portions displayed a very low signal intensity on T2W without enhancement, suspicious of a hemorrhagic component. There was another lobulated, intraconal nodule at the right orbital apex with homogeneous enhancement (arrow head)



**Figure 5:** (a) Hematoxylin and eosin staining revealed collapsed, thin-walled, vascular channels (\*) associated with organized thrombus. (b) CD31 staining highlighted the endothelial cells of the collapsed lumen (arrow head). (c) Smooth muscle actin (SMA) staining highlighted the smooth muscle cells in the vascular wall

Multiple CVMs and bilateral orbital CVMs are rare. This patient had a bilateral orbital CVM and another CVM on the left cheek. In this case, the lesion was at the orbital apex, which is a rare location for a CVM. The acute onset of the symptoms in this patient related to an intralesional hemorrhage in the CVM. The expansion of the intralesional hemorrhage in the CVM compressed the optic nerve in the very restricted space of the orbital apex, causing an acute visual loss and orbital pain.

An acute intralesional hemorrhage of an orbital CVM is rare,<sup>[3]</sup> with only a few cases having been reported.<sup>[4-7]</sup> Interestingly, most of the acute lesions were in the left eye.<sup>[4-7]</sup> The mechanism of intralesional hemorrhaging of an orbital CVM remains unclear. It may be caused by inflammation of the small capillaries within the CVM. this would obstruct the blood circulation and thereby induce a hemorrhage.<sup>[6]</sup> However, a study by Rootman *et al.* revealed very little inflammation in the histologic features of intralesional hemorrhages, and they postulated that the cause of the thrombosis was a vascular stasis eddy formation that induced hypercellularity and stromal changes.<sup>[2]</sup> As the pathology investigation of our patient did not reveal the presence of inflammatory cells, we concluded that the cause of the thrombosis in our case was slow hemodynamics in the CVM.

The asymptomatic orbital CVM in the right orbit of this patient did not need surgical treatment. The CVM may have been related to the blue rubber bleb nevus syndrome.<sup>[8]</sup> Our patient was investigated for other CVMs in the body; fortunately, none were found.

An orbital CVM can present with an acute and painful visual loss that mimics another orbital disease, idiopathic orbital inflammation. A careful history and examination, including a thorough review of orbital imaging, should result in a correct diagnosis and proper management.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot bechrological order guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

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