

## CASE REPORT

**VISUAL IMPROVEMENT FOLLOWING THE REMOVAL OF CHRONIC ORBITAL CAVERNOUS HEMANGIOMA****Raisha Pratiwi Indrawati<sup>1</sup>, M. Rinaldi Dahlan<sup>1,2</sup>**<sup>1</sup>*Department of Ophthalmology, Faculty of Medicine, Universitas Padjadjaran, Bandung, Indonesia*<sup>2</sup>*National Eye Center, Cicendo Eye Hospital, Bandung, Indonesia**Email: raishapi@gmail.com***ABSTRACT**

**Introduction:** *Orbital cavernous hemangioma is a benign, well-defined vascular lesion, located mainly in the intraconal space. Surgical treatment is indicated in symptomatic cases, such as proptosis or visual disturbance. This study aims to report the visual improvement of the successfully-managed chronic orbital cavernous hemangioma.*

**Case Report:** *A 51-year-old man came to Cicendo Eye Hospital with chief complaint of protrusion of the right eye for seventeen years, accompanied by blurred vision two years earlier. Ophthalmological examination revealed decreased visual acuity, together with ocular movement restriction and relative afferent pupillary defect. Computed tomography (CT) scan showed a retrobulbar mass, attached to the lateral and medial rectus muscle, and shifting the optic nerve superiorly. The tumor was removed through inferior anterior orbitotomy with a transconjunctival incision, and the histopathological finding revealed a cavernous hemangioma. Follow-up examination showed improvement in the patient's condition, measured by no protrusion remaining and the achievement of best-corrected visual acuity (BCVA) of 1.0.*

**Discussion:** *In the case of a benign intraorbital tumor, the orbitotomy approach should be best selected based on its anatomical location and involvement of adjacent structures. Despite the size of the tumor, chronic duration, and disturbing manifestation, improvement of visual acuity and structural appearance could be achieved if the tumor is removed correctly and the adjacent structure has not been severely damaged.*

**Conclusion:** *Cavernous hemangioma is a benign lesion in which surgical treatment is indicated in symptomatic cases. The right approach of orbitotomy is needed to give a good outcome anatomically and functionally.*

**Keywords:** *cavernous hemangioma, anterior orbitotomy*

**INTRODUCTION**

**C**avernous hemangioma, also known as cavernous vein malformation, is the most common orbital lesion in the fourth to fifth decade of adulthood, with 60% of cases occurring in women. Cavernous hemangiomas consist of a proliferation of large endothelial-lined vascular canals surrounded by a well-defined fibrous capsule. This is a benign tumor with slow progression, with clinical appearance varying from asymptomatic to manifest in the form of proptosis, decreased visual acuity, or double vision, and can occur unilaterally or bilaterally.<sup>1-3</sup>

The diagnosis of cavernous hemangioma can be established from clinical manifestations or incidental imaging findings without previous signs and symptoms. Approximately 30% of cases of cavernous hemangioma have no symptoms and do not require immediate invasive treatment. Treatment in the form of surgery can be considered in conditions accompanied by symptoms, especially those involving impaired visual function. The surgical technique chosen can depend on several things, including imaging findings in the form of the size and location of the lesion as well as the preferences of the operator.<sup>2,4,5</sup> This case report aims to discuss the management of patients with orbital cavernous hemangioma.

### CASE ILLUSTRATION

A 51-year-old man came to the Reconstruction, Oculoplasty and Oncology Polyclinic (ROO) of the National Eye Center (PMN) Cicendo Eye Hospital with complaints that his right eye appeared enlarged about seventeen years before presentation. Complaints were felt to be slowly worsening, accompanied by the rise upward of the right eyeball. The patient also had a history of blurry vision two years earlier, without any history of double vision. History of systemic disease, ocular trauma, ocular surgery and previous medication was denied. There was no history of similar complaints in the family.

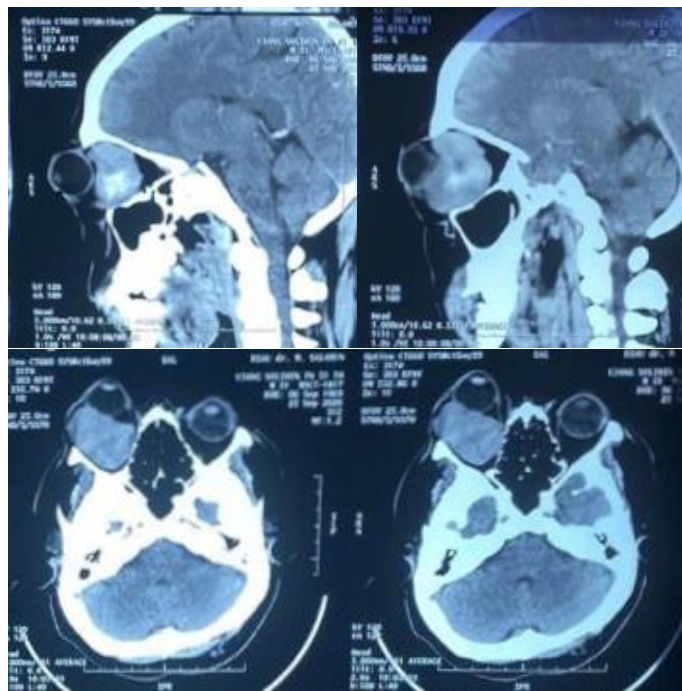


**Figure 1.** Clinical picture of the patient preoperatively (upper) and postoperatively (lower). Upper: The right eye shows proptosis with a shift to the superior as seen in anterior view and lateral view. Lower : Improvement of the proptosis 1 month after surgery.

General status examination was found within normal limits. Ophthalmological examination showed close-to-face finger counting (CFFC) visual acuity in the right eye and 0.25 in the left eye. The position of the right eye was hypertropia. The movement of the right

eye was limited in all directions, while the movement of the left eyeball was normal. Intraocular pressure by palpation was normal in both eyes. Examination of the lid of the right eye showed proptosis without lagophthalmos, with a palpable mass measuring  $\pm 1.5 \times 1.5$  cm superiorly and  $\pm 2.5 \times 1.5$  cm inferiorly accompanied by hyperemic conjunctiva. Biomicroscopic slit lamp examination showed normal anterior segment. The direct light reflex in the right eye was decreased with preserved indirect light reflex, showing grade III relative afferent pupillary defect (RAPD). Fundus examination in the right eye showed a slightly pale optic nerve head, whereas, in the left eye it was within normal limit.

The results of the CT scan showed a mass in the right retrobulbar area with a size of  $\pm 4.1 \times 2.6 \times 4.3$  cm, especially on the lateral side, which seemed to be attached to the lateral and medial rectus muscles, and pushed the optic nerve superiorly, with inhomogeneous enhancement. The patient was then diagnosed with proptosis of the right eye due to retrobulbar mass, suspected hemangioma with differential diagnosis of meningioma and Schwannoma, along with compressive optic neuropathy.



**Figure 2.** CT scan of the head and orbit shows a mass in the retrobulbar area that pushes the eyeball anteriorly and superiorly

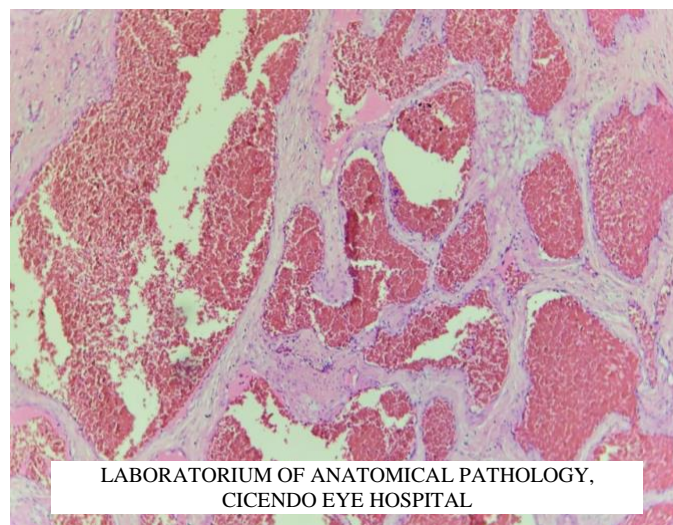
The patient then underwent a mass debulking surgery, using anterior inferior orbitotomy with a transconjunctival approach which was performed under general anesthesia. Intraoperative finding showed encapsulated tissue measuring  $5.8 \times 3.5 \times 2.5$  cm, brownish-red in color with a rubbery consistency, which was suggested as hemangioma. The patient was given intravenous therapy of methylprednisolone and tranexamic acid, as well as oral ciprofloxacin,

mefenamic acid and antibiotic eye ointment in the right eye. One day after surgery, ophthalmological examination of the right eye revealed visual acuity of 1/60 in the right eye with limited eye movement, but there was no more proptosis.



**Figure 3.** The intra-operative findings revealed a well-defined, encapsulated, violaceous-red mass, suggestive of a hemangioma.

Ten days following surgery, the patient came for a follow-up visit, and the visual acuity of the right eye was 3/60 pinhole 0.125 and in the left eye was 0.4 pinhole 0.63. There were still limitations of eye movement to temporal, superotemporal, and inferotemporal directions in the right eye. The histopathological result revealed a cavernous hemangioma without any sign of malignancy. A month later the patient came again and his best corrected visual acuity was improved to 1.0 with no protrusion left.



**Figure 4.** Histopathological finding showed large vessel proliferation, revealed the diagnosis of orbital cavernous hemangioma.

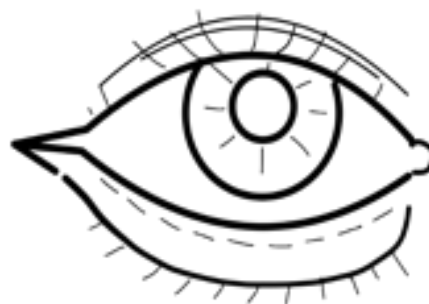
## DISCUSSION

Cavernous hemangioma is a non-infiltrative vascular benign tumor consisting of proliferation of large vascular canals with slow progression and generally occurs unilaterally. According to the International Society for the Study of Vascular Anomalies (ISSVA) classification, currently cavernous hemangioma is classified as a malformation, hence, it can be referred to as a cavernous vein malformation. Cavernous hemangiomas can occur from birth but often lie dormant for years without any clinical signs or symptoms. The main manifestation of cavernous hemangioma is proptosis which is generally axial, with slow progression and without pain. Other complaints include limitation of eyeball movement and diplopia in cases of larger tumors and can be accompanied by a decrease in visual acuity due to compression of the optic nerve or hyperopia.<sup>1,3,6</sup> The patient in this case report came with complaints of bulging eyes that have been felt for seventeen years, showing slow tumor progression which is suggested as a benign tumor. Patients also felt a decrease in visual acuity without double vision. Ophthalmological examination of the patient revealed a suspicion of compression of the optic nerve which exacerbated the patient's complaints.

Ancillary testings play an important role in diagnosing tumors, whether symptomatic or not, and differentiating the possibility of benign or malignant lesions. These examinations will also be useful in determining the approach to be used in surgery. These tests include CT scan, Magnetic Resonance Imaging (MRI), and histopathological examination. Cavernous hemangioma on CT scan will appear as a well-demarcated, homogeneous, oval-rounded mass with a higher density than muscle. Pathognomonic signs of a hemangioma include phlebolith calcification, bony hypertrophy, or intraosseous extension. Lesions can be seen in the intraconal space or extend extraconally. T1-weighted (T1W) MRI examination can show isointense homogeneity to muscle and cortical gray matter and hypointense to fat, whereas T2-weighted (T2W) produces hyperintense images to fat and brain. Staining with gadolinium may show a homogeneous ring. Histopathologically, a cavernous hemangioma is a lesion that forms from a cavernous vascular space lined with endothelial cells with connective tissue septa, which may be partially or completely filled with blood. This tumor is covered by a pseudocapsule which will be passed by the feeder vessel.<sup>6-9</sup> The ancillary examination performed in this patient was a CT scan, where the results of the CT scan showed the presence of a well-defined, round oval retrobulbar mass, and located in the anterior 2/3 orbit, accompanied by calcification. The mass was seen to push the eyeball anteriorly and superiorly causing proptosis in the patient, and it appeared to be attached to the muscle and pushing on the optic nerve, explaining the clinical findings obtained on ophthalmological examination. The results obtained from this CT scan are

useful in estimating the working diagnosis and differential diagnosis, they also play a role in determining the approach to be used for tumor removal. Establishing a definitive diagnosis can be made with a histopathological examination, which is useful for determining further treatment plans and the prognosis of the patient.

The management of cavernous hemangiomas can be either surgical or non-surgical. Asymptomatic cases can be observed first because they have slow progress and rarely cause complications. Surgery is an option when the tumor causes symptoms such as disturbing proptosis, visual disturbances, or double visions. The technique used for tumor removal can be anterior, lateral, or transcranial orbitotomy, depending on the anatomical location and relationship to the surrounding orbital structures. Anterior orbitotomy is currently the most widely used technique for lesions of the anterior 2/3, both extraconal and intraconal that do not involve the apex. This technique can be used for both incisional biopsy and total excision of the mass. Several anterior orbitotomy approaches include the transconjunctival, transcutaneous, and transcaruncular approaches. The incisional approach is chosen based on the technique that allows for good access to the mass, with minimal risk to the patient. Complications that may occur with anterior orbitotomy include bleeding, palpebral hematoma, mydriasis, loss of vision, or compartment syndrome. Deeper tumors can be removed by lateral orbitotomy technique, whereas tumors involving the apex must be accessed by craniotomy.<sup>2,7,10</sup> CT-scan results showed intraconal lesion, mostly lying inferiorly and pushing the eyeball superiorly. This description became the basis for performing an anterior orbitotomy with a transconjunctival approach in the inferior section. The steps taken for the conjunctival incision were by firstly performing a lateral canthotomy and inferior cantholysis, then retraction of the palpebral margin, followed by a deeper incision until it reached the inferior orbital rim, then a periosteum dissection was performed. In this case, after the mass was reached, further exploration and complete removal of the mass was carried out, and a purplish-red encapsulated mass was obtained, suggestive of cavernous hemangioma.



**Figure 5.** Illustration of inferior anterior orbitotomy technique with transconjunctival approach

Cavernous hemangioma is a benign tumor, therefore it often has a good prognosis. Visual acuity can improve after surgery, although in some conditions hyperopia or other complications can persist. However, recurrence can occur in conditions of incomplete tumor excision.<sup>3,6,11</sup> The patient in this case showed an improvement both structurally and functionally. The proptosis was resolved and visual acuity was much improved compared to the initial condition before surgery. Despite the size of the tumor, chronic duration, and disturbing manifestation, eye structure and function could be much improved if the tumor is removed correctly and adjacent structure has not been severely damaged. Long-term follow-up is needed to prevent the recurrence of the condition.

## CONCLUSION

Cavernous hemangioma is a benign vascular tumor with slow progression but often requires surgical management if it has clinically significant manifestations. The appropriate orbitotomy technique should be decided based on clinical and ancillary examination considerations. Anterior orbitotomy is one of the surgical techniques that can be performed in the treatment of intraconal and extraconal cavernous hemangioma, especially for those located in the anterior 2/3, which will give good surgical results anatomically and functionally.

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