CASE REPORTS

# Orbital cavernous hemangioma

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## Abstract

A case of orbital cavernous hemangioma (OCH) in the Third Affiliated Hospital of Inner Mongolia Medical University was collected and analyzed on the basis of diagnosis, physical examination and treatment. Misdiagnosis of OCH is very common since it is a rare disease. So this paper aims to enhance the doctors' awareness of OCH during clinical practice.

**Key Words:** Orbital cavernous hemangioma, Lacrimal gland area, Retina, Cataract

## 1 Medical record

### 1.1 General information

A 64-year-old female was admitted to our hospital due to "pain in the right eye for 4 years and proptosis, diplopia with decreased visual acuity for 2 years" on June 29, 2010. The patient suffered from pain of unknown origin in the right eye, with headache 4 years ago. The syndrome she had two years ago, such as right eye proptosis, eye pain, orbital pain with headache, diplopia with decreased visual acuity while the eye turning left and upward, significantly progressed without redness, increase in secretions, or fearless light tears during the past two months. Her past medical history included hypertension of 1 years' duration.

### 1.2 Physical examination

Data on the physical examination revealed her temperature 36.5°C, pulse 70 beats per minute, breathing 20 per minute, blood pressure 160/90 mmHg. The general condition of the whole body seemed fine. Ophthalmic testing result showed oculus dexter (OD) 0.5 (corrected 0.7) oculus sinister (OS) 1.0. The signs and synptoms included: mild right eyelid ptosis, the upper eyelid cover above Corneal limbus 3 mm, width of palpebral fissure 6 mm, eye shift downward 5 mm. Left eye upward movement is limited, no hyperemia in bulbar conjunctiva, transparent cornea, normal pupil size, normal reflex, phacoscotasmus and vitreous opacity. Eye ground examination results showed that optic nerve head color was normal with distinct edges. An unpushable mass is palpable in the right orbital edge without tenderness, exophthalmos 17 mm (right eye), and 12 mm (left eye), orbital distance 102 mm. Orbital tensions are T+2 (right eye) and Tn (left eye). Both hirschbeng and anterior segment of the left eye were normal.

### 1.3 Auxiliary examination

Orbital CT scan result included: an oval soft tissue density was visible on the upward side of right rectus, CT values were 52Hu, the maximum ranged about 2.2 cm × 1.6 cm × 1.4 cm in size with fine edge, the rectus and the eye were oppressed (see Figure 1). Eye ultrasound examination revealed that: a hypoechoic mass (2.6 cm × 2.1 cm) with clear boundary was detected above the right orbit, whose flow signals within the mass could not be shown, and the venous blood flow presented with low-speed signals (see Figure 2).

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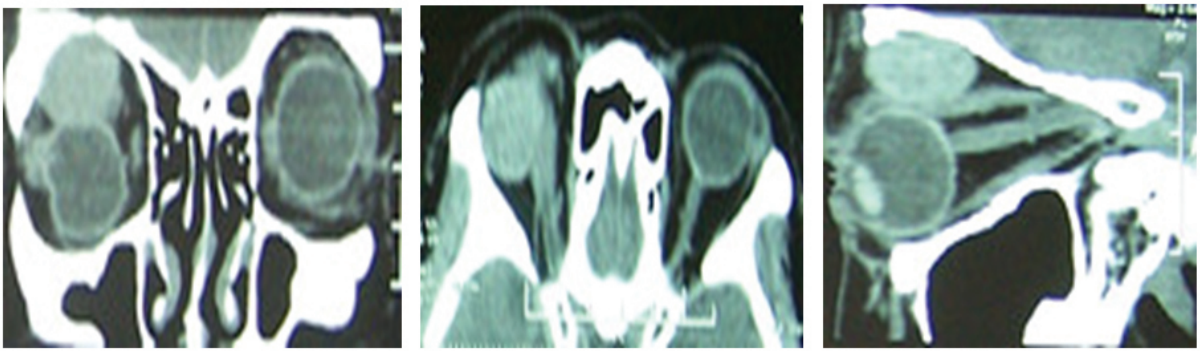

**Figure 1:** Preoperative CT examination

### 1.4 Primary diagnosis

(1) Orbital lesions in the right eye
(2) Right ptosis
(3) Refractive error of the right eye
(4) Binocular senile cataract
(5) Vitreous opacities of both eyes
(6) Hypertension grade 2

### 1.5 Treatment

The patient underwent extra relevant examination after admission. We would prefer anterior skin incision as surgical approach under general anesthesia. The surgery started with upper outwards orbital margin to open the orbit, and the top of the outer periosteal then was cut apart, with partial delamination. Partial bones around orbital margin bones were removed with rongeur so that the tumor was visible. It is a prunosus round tumor, 25 mm diameter in size, with intact capsule in the lacrimal area. The tumor was gently stripped and took out (see Figure 3).

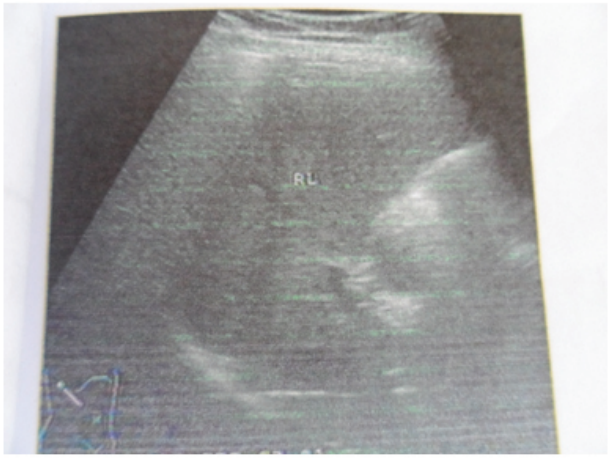

**Figure 2:** Preoperative ultrasound examination

The incision was stitched if no residual tumor tissue was detected, which indicated that the surgery was over. The excised tissue was sent for pathology examination and was tested to be orbital cavernous hemangioma (see Figure 4). Postoperative treatment of anti-inflammatory, hemostasis, infection prevention was performed with good outcome. The patient had no protopsis, and eye movement was normal, without other discomfort during two years' follow-up.

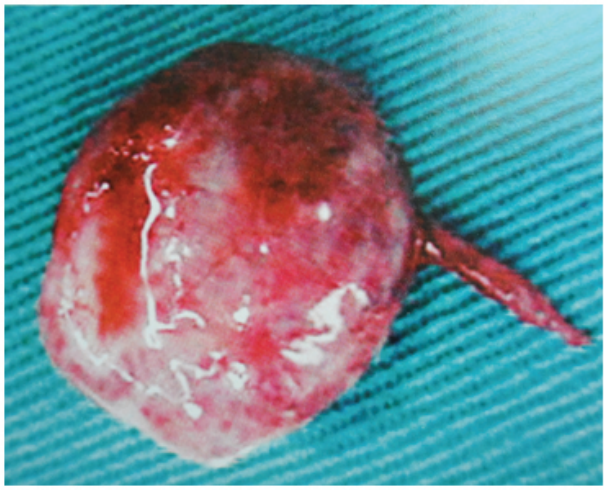

**Figure 3:** The tumor

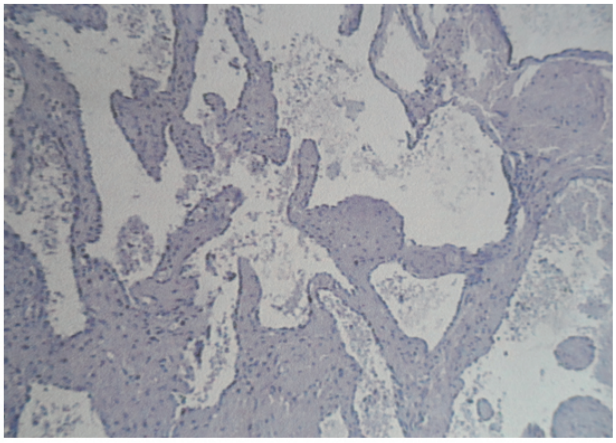

**Figure 4:** Pathology examination

### 1.6 Confirmation of the diagnosis

(1) Orbital cavernous hemangioma in the right eye
(2) Right ptosis
(3) Refractive error of the right eye
(4) Binocular senile cataract
(5) Vitreous opacities of both eyes
(6) Hypertension grade 2

## 2 Discussion

### 2.1 Dr. Chengkun An

*Dr. Chengkun An is the doctor at The Third Affiliated Hospital of Inner Mongolia Medical University.*

Orbital cavernous hemangioma (OCH) is the most common benign orbital tumor. It may be difficult to differentiate from other intraconal lesions, so that its pathogenesis is uncertain. It is a vascular malformation characterized by the presence of sinusoids with fine walls, usually observed in adults and thought to be benign, accounting for 14.5%-21.3% of primary expending masses.[1–3] Histopathology showed that tumor was encapsuled, whose capsule was thicker than those in the rest of the body, composed of vessel lomen with intimal hyperplasia, and loose distribution of smooth muscle in the blood vessel wall and matrix. OCH is low-flow hemangioma in hemodynamics and histology.[4] The management in case of OCH mainly depends on surgery, based on pathology imaging finding, due to its unique characteristics.

The causes of the disease in this case are uncertain. Scholars used to believe that the tumor was evolved from capillary hemangioma, the pressure within the lumen increased, accompanying with high degree of expansion, thus forming blood vessels sinus. Clinical manifestation and histopathology, however, do not support this argument. Cavernous hemangioma, a kind of hamartoma, is formation of a variety of cellular components of tumor, more mature than capillary hemangioma, develops along the vascular, with smooth muscle cells in the vessel wall. The tumors were round or oval with intact capsule, sliced dark red. The cavernous sinus cavity was filled with blood, whose compartment consisted of connective tissue and smooth muscle cells, endothelial cells hyperplasia.

### 2.2 Dr. Bin Shen

*Dr. Bin Shen is the doctor at The Third Affiliated Hospital of Inner Mongolia Medical University.*

Clinically, the appearance of signs and associated symptoms include: (1) Most OCH lesions are deep orbit or muscle cone, whose most presence of long-term proptosis. A soft and smooth mass with clear boundary is palpable without tenderness if the lesion is before orbital or periorbital area. (2) There is no specific symptom if the tumor is small or at the deeper parts. Treatment is usually taken when the physical examination or head CT revealed occupying orbit. (3) Progressive vision loss. The tumor presses posterior pole of eyeball, causes axial shortening, which may further lead to hyperopia and astigmatism. Primary optic atrophy can result from long-term pressure on optic nerve if the lesion is at orbital apex. (4) The symptom of diplopia may occur in case the tumor is too large that causes limited eye movement. (5) Orbital pressure is much associated with volume and lesions, raging from (+)-(++). Elastic resistance could be felt while pressing the eyeball.

The patient had mild ptosis in the right eyelid before admission. The other symptoms include: the upper eyelid cover above corneal limbus 3 mm, blepharophimosis width 6 mm, the right eyeball shift downward 5 mm, compared with the left eye, upward and left movement is limited. An unpushable mass is palpable in the right orbital edge without tenderness, exophthalmos 17 mm (right eye), and 12 mm (left eye), orbital distance 102 mm. Orbital tensions are T+2 (right eye) and Tn (left eye). The clinical symptoms and signs are consistent with the clinical manifestations of cavernous hemangioma.

### 2.3 Dr. Lili Gao

*Dr. Lili Gao is the director of Department of Ophthalmology at The Third Affiliated Hospital of Inner Mongolia Medical University, specializing in cataract and oculoplasty.*

B ultrasound imaging of cavernous hemangioma is more typical, showing round or oval mass with more and evenly distributed echo, sound attenuation[5] and slightly compressible. The tumor may be compressible, which is bound up with tumor location within the orbit. If the tumor is located at the front, compressibility will be evident, if not, it is not obvious. Color doppler ultrasound caught the sign, that the tumor is lack of blood flow as the blood flow within the sinus was extremely slow. However, color doppler ultrasound could not delineate a good spatial location of the tumor due to its limited level of each frame, so its role as location diagnosis is limited.

CT is excellent for locating the mass within the orbit, and coronal plane is useful to reveal the relationship between the tumor and the optic nerve. In addition, whether the tumor has progressed to the brain could be determined by the appearance of transparent triangle through orbital apex since tumors that originated in the orbital apex or spread back are usually lack of such signs.[6] The way that CT detects the tumor depends heavily on density difference of tumor tissue so that localization diagnosis of cavernous hemangioma and extraocular muscles and other density remains challenging due to the homogeneous density of the tumor, and there is no big difference between cavernous hemangioma and other benign solid tumors. Though the clinical diagnosis could be made through the analysis of its site and shape, it is not con-

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ducive to localized and qualitative diagnostic in case of similar tissue density. Small circular high density calcification could be seen in some tumors, as well as in lymphangioma, capillary hemangioma and other tumors, whose essence is stone of vain, so calcification is believed to be of no value in identifying the tumor. Dynamic CT scan showed signs of gradual strengthening, which were relatively specific signs in the diagnosis of cavernous hemangioma. The decrease in density is adverse to our observation of the exact scope of lesions.

MRI serves as a useful tool to provide multi-planar and multi-parameter imaging of the soft tissue with high resolution and precise positioning, whose function to reveal tumor's relationship with optic nerve is advantage over CT.[7] MRI showed that the mass produces low signal intensity on axial T1 weighted image (T1WI) and a homogeneous and markedly high signal intensity on axial T2 weighted image (T2WI), which also could be found in other tumors such as neurofibromas, hemangiopericytomas, nerve sheath tumors and lymphatic tumors, thus, the differential diagnosis is of limited value. T2WI high signal is reported to increase along with the extension of time of echo,[8] which is detected in lesions of rich liquid, such as lymphangioma, cystic nerve sheath tumors and cysts, while T2WI signals for other orbital benign tumors does not change. The difference in the resonance frequency of cavernous hemangioma and orbital fat-rich liquid was large, and chemical shift artifact was formed around the tumor. Whether the performance could hint of the disease requires further studies. Progressive strengthening during enhanced scanning serves as a relatively specific signs in the diagnosis of the disease[9, 10] so that MRI enhancement scanning advantages over CT: the longer MRI contrast agent stays in the tumor, the more sensitive to paramagnetic MRI contrast agent, showing a clear progressive strengthening process. It is obvious that MRI is better than CT on localized and qualitative diagnosis, so that cost for MRI is higher.

Orbital CT scan results including: an oval soft tissue density was visible on the upward side of right rectus, CT values were 52Hu, the maximum ranged about 2.2 cm × 1.6 cm × 1.4 cm in size with fine edge, the rectus and the eye were oppressed. Eye ultrasound examination revealed that: a hypoechoic mass (2.6 cm × 2.1 cm) with clear boundary was detected above the right orbit, whose flow signals within the mass could not be shown, and the venous blood flow presented with low-speed signals. Auxiliary examination of patients: Eye color Doppler ultrasound and CT results were consistent with orbital cavernous hemangioma features.

### 2.4 Dr. Xinping Kong

*Dr. Xinping Kong is the Deputy Chief Physician of Department of Ophthalmology at The Third Affiliated Hospital of Inner Mongolia Medical University, specializing in Diseases of the retina and Orbital diseases.*

OCH is a common benign tumor, which may occur in any part of the orbit, commonly found within muscle cone, and clinically presented as progressive painless proptosis. It often causes vision loss when the orbital apex is oppressed. Intraconal OCH should be distinguished from the following retrobulbar tumors with capsule: (1) optic nerve glioma; (2) meningioma; (3) schwannomas. Optic gliomas mostly occur in young children, characterized by earlier impaired vision, exophthalmos, papilledema or atrophy. CT imaging shows conical enlargement, or circular swelling of thicken front and thin back along the optic nerve, involving optic foramen and cranium. Meningiomas occur in middle-aged women over the age of 40, characterized with vision loss, papilledema, and optic nerve atrophy secondary to ciliary vessels, visual impairment after prominent. B ultrasound reflects less echo of the tumor, and attenuation with incompressible. CT scan shows tube, spindle-shaped thickening or conical mass. MRI reveals that the tumor derives from optic nerve sheaths, spreading within tube or intracranial area. Schwannomas may occur in any nerve within the orbit other than the optic nerve. Tumor shadow is mostly located outside the muscle cone, with clear edge and density heterogeneity, sometimes with tenderness, cystic necrosis. B ultrasound reflects less echo, and strong transparent sound. CT value is low during CT scan, whose degree of enhancement after injection enhancer generally is no more than 20Hu. MRI finds cranio-orbital tumors when the lesions are at orbital apex. Extraconal OCH should be identified from venous angioma, benign pleomorphic adenoma of the lacrimal gland. Venous angioma occurs earlier than cavernous hemangioma, is often found among children and teenagers, located in the orbital quadrant, and presented with proptosis and inferolateral shift. Proptosis is much associated with position, for example, the symptom of Proptosis is obvious when the patient bow or internal jugular vein is oppressed. It relieved when the patient is standing. B ultrasound demonstrates occupying lesions with irregular shapes, unclear or unsmooth boundary, varying amounts of echo, and multiple tubular or sheet-shaped echo-free zone. In addition, some scholars believe that there is no calcification in cavernous hemangioma, while calcification and phlebolith are mostly seen in venous angioma.[11] Pleomorphic adenoma (benign mixed tumour) of lacrimal gland usually originates in the lacrimal fossa, manifested as significant deformation of the eyeball due to great pressure from the tumor, whose B ultrasound reflects moderate echo. CT found bone resorption in the lacrimal gland. The patient was diagnosed as cavernous hemangioma according to her clinical manifestation and radiological imaging, while its occurrence in the lacrimal gland is rare.

### 2.5 Dr. Xiangyang Xin

*Dr. Xiangyang Xin is the Deputy Chief of Department of Ophthalmology at The Third Affiliated Hospital of Inner*

*Mongolia Medical University, specializing in orbital diseases, cataract and ocular trauma.*

Surgical resection and close observation are the main choice for OCH treatment. Follow-up visits are feasible for tumors of small volume with atypical syndrome due to its slow progressive growth.[12] Surgery, however, is required clinically in most cases of delayed treatment on clinical signs and symptoms. An appropriate surgical approach depends on a comprehensive preoperative assessment of tumor nature, location, and the extent of adhesions. We believe that anterior orbitotomy could be performed for tumors inside, outside or above muscle cone, intramuscular optic nerve according to our accumulated experience in clinical practice during recent years. Tranconjunctival approach seems an ideal choice for upward, outward and downward side of outside of the optic nerve, above the optic nerve, within the upper side of smaller volume. It also shortens operative time and causes less damage without scars, which significantly reduced the incidence of complications such as ptosis while comparing with lateral orbitotomy and anterior orbitotomy. Compared with the transconjunctival fornix, it could reach directly into the muscle cone without a second surgical space involved, thus avoiding the extrapyramidal fat prolapse and the destruction of intramuscular membrane, and reduce the tissue damage. Conjunctival approach is an available treatment for the vast majority of cavernous hemangioma, therefore, it should be regarded as a primary choice for the management of the disease. Lateral orbitotomy shall be employed for tumors with heavier adhesion in the orbital apex.[13] The surgery is usually operated on sawed orbitotomy orbital wall, with strong postoperative reaction form the tissue, but bleeding could be easily handled without causing damage to eye muscles or affecting vision during the operation due to a good exposure of the surgical field. Complete resection of the larger tumor could be obtained as well.

It is rare when the lesion is located in the lacrimal gland in the case, whose preoperative clinical and radiological diagnosis initially identifies the symptoms as cavernous hemangioma. Therefore, the patient underwent anterior orbitotomy for surgical resection, staring from top of the outer skin incision into the orbit, then eyebrow lower edge as surgical incision to expose the tumor. She recovered without recurrence during two years' follow-up.

## 3 Conclusion

In summary, cavernous hemangioma at the lacrimal gland is rare so that it could be easily misdiagnosed. The paper aims to improve doctors' understanding of the disease, expand their clinical thinking and improve clinical diagnosis and treatment of rare diseases.

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