Case Report

Multiple heterogeneous tumors in orbit: a case report

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Abstract: Primary orbital tumors are common; clinically common are single tumors in orbit; and cavernous hemangioma is the most common. Multiple tumors in the same orbit are rare, which have homology and heterogeneity. Multiple tumors of homology are multiple textures of a tumor while multiple heterogeneous tumors have two or more textures. Various tumor tissues, heterogeneous multiple orbital tumors are sporadic. Our department admitted one patient in October 2015. A 54-year-old male patient had proptosis growing gradually in his right eye beginning ten years ago, with decreased vision for more than one year. The primary diagnosis of the right eye orbital double-source primary tumor was diagnosed by imaging. The pathological examination after the operation was confirmed as cavernous hemangioma and schwannomas.

Keywords: Orbital tumor, cavernous hemangioma, schwannomas

Introduction

Multiple orbital tumors are rare diseases. Most multiple tumors are homologous tumors, such as multiple cavernous hemangiomas. While multiple orbital heterogeneous tumors are rarely reported. This patient has multiple tumors of the right orbit, which are derived from cavernous hemangioma and schwannomas.

Cavernous hemangioma is the most common orbital tumor, accounting for 18.09% of orbital tumors [1]. In the incipient stage, cavernous hemangioma without compression nerves and surrounding muscle tissue can have no obvious clinical symptoms, so patients often find tumors due to other reasons for imaging examination. Orbital schwannomas are a common benign tumor, which incidentally is the fourth in the benign orbital tumors [2]. Schwannomas are benign tumors that originate from Schwann cells. They occur mostly in the intracranial and peripheral nerves. Most of them originated in the orbital area. Patients often come to the hospital for reasons such as eyeball protrusion, eye movement disorder, and decreased vision. We reported this rare presentation of multiple orbital heterologous tumors.

Case report

A 54-year-old male patient presented to the ophthalmic clinic with proptosis, eye movement disorder, and vision that had gradually decreased during the past ten years. This patient was not provided with proper attention and targeted treatment due to family poverty and absence of pain in the right eye (OD) (Figure 1A). The exophthalmos was measured as 28 mm of the right eye and 15 mm of the left eye (OS). The patient’s diseased eye had 13 mm proptosis compared to the left. Naked visual acuity was blindness for the right eye and 0.8 for the left. The OD can confirm that the intraorbital pressure is much higher than the data from normal people, but the intraocular pressure in the OD was normal. Both the intraocular and intraorbital pressure in the OS was normal.

Magnetic resonance imaging revealed a 1.7 cm×2 cm×2.5 cm and a 3.8 cm×3.5 cm×4 cm well-defined intraconal mass in the anterior and posterior right orbit (Figure 2). The tumor in the anterior orbit hypointense on T1-weighted images (T1WI), homogeneously hyperintense on T2-weighted images (T2WI), without contrast enhancement. The other one located in the posterior orbit was isointensity on T1WI, iso-
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Figure 1. Right eye proptosis was identified before the operation (A). After the surgery, the proptosis of the right eye had disappeared for seven days (B).

Figure 2. MRI revealed two well-defined pieces of orbital mass in the right orbit.

tensity combined with hyper-intensity on T2WI, and there was increased signal intensity in contrast enhancement (Figure 2). It was observed that the inner bone wall of the right orbit was obviously thinner by computed tomography (CT) scan (Figure 3). Based on the above clinical and radiological findings, a preoperative diagnosis of benign intraorbital neoplasm was made, and surgical excision was planned.

The right lateral canthal was cut open, and the lateral skin horizontal incision was extended to the temporal side by about 2.5 cm. The tissue was carefully separated and the upper bone of the orbit was removed by a bone saw. The gray-white tumor was seen on the outer part of the orbit. The surface was smooth and the capsule was intact. It was considered as a schwannoma. It was considered to be a cavernous hemangioma from the outer rectus and the inferior rectus muscle to the muscle cone. See the larger color purple red tumor tissue, showing a round shape, clear boundary, and intact capsule (Figure 4).

Histopathological examination of the specimen revealed the tumor’s tissue structure. The tumor, which was located in the anterior orbit, had cells that were spindle shaped and were arranged in fascicles along with Antoni A-pattern structures and loose paucicellular Antoni
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Discussion

Cavernous hemangioma is the most common primary tumor in the sputum in adults, and women are more common than men. Schwannoma, a Schwann cell is originating from the peripheral nerve sheath, which grows slowly and has clear boundaries with surrounding tissues. They are often benign. The intraocular nerve schwannomas account for 1% to 4% of the primary tumors in orbit [3]. There is no gender difference in the incidence of schwannomas, which can occur at any age, and often occurs in middle-aged people. The cavernous hemangioma and schwannomas occurred in the same orbit are very rare; this case should be the first case report.

Due to the slow growth of cavernous hemangioma and schwannomas, there are often no obvious clinical symptoms in the early stage. The cavernous hemangioma in orbit is different from other cavernous hemangiomas, and there is no spontaneous bleeding, which increases the confusion. As the tumor volume increases, the eyeballs protrude; the surrounding extraocular muscles are pressed, causing eye movement disorders; the oppressive nerves cause vision loss. The tumor in the apex of orbit tends to have symptoms of decreased vision at an early stage. The patient’s medical history lasted for more than ten years, and the slow and gradual course of disease often caused the patient to ignore the progress of the disease.

Cavernous hemangioma and schwannomas can often be identified preoperatively by imaging techniques, such as MRI, CT, and ultrasound. CT imaging of cavernous hemangioma is mostly round and elliptical, mostly located in the muscle cone, squeezing the bone wall, causing deformation of the orbital wall, but often without bone destruction. After the injection of contrast medium, progressive enhancement signs appear, suggesting that the lesion lacks nourishing blood vessels [4, 5]. It was confirmed by a large amount of experimental data that ultrasound with 15 MHz showed that the cavernous hemangioma was the clearest, with a rounded shape with a clear boundary-the slow or medium acoustic absorption. The slow

B-pattern structures, the latter occupying the majority of areas (Figure 5A). Based on these observations, a diagnosis of schwannoma was made. Positive expression of S-100 protein and Ki-67 3% positive supported the diagnosis (Figure 5B and 5C). The other which was located in the posterior orbit had confirmed that it was a typical tissue of the cavernous hemangioma (Figure 6).

Postoperatively, the patient had a good cosmetic result and has remained asymptomatic over a 4-year follow-up period. To our knowledge, this is the first reported case of such lesions in orbit (Figure 1B).
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The blood flow of the point arteries in the cavernous hemangioma shows in CDI, but it is not diagnostic [6-9]. Cavernous hemangioma, the tumor is an isointensity on T1WI, the signal is lower than fat, similar to the extraocular muscle. Tumors were hyperintense on T2WI, and the signal was significantly enhanced after injection of Gd-dTPA. Tablet or all tumor fillings can be seen using contrast medium and fat suppression techniques [5, 10, 11].

The shape of schwannomas is more complicated depending on the growth mode, and it is mostly located in the posterior segment of the orbit, which is related to nerve distribution. The patient's schwannomas were located in the anterior orbit around the lacrimal gland, less oppression of the optic nerve, and peripheral extraocular muscles; so the patient did not appear to have obvious eye movement disorders or decreased vision in the early stage. In the CT scan of schwannomas, the density was close to the optic nerve and extraocular muscles, but lower than the cavernous hemangioma. We can find a weak echo, strong sound permeability by the B-ultrasound. It can be seen by CDI that is the enrichment blood flow signal, can be distinguished from cavernous hemangioma. Through MRI, we can get a hypointense on T1WI, and hyperintense on T2WI, and images with different signal intensities are useful for observing the positional relationship between the tumor and surrounding tissues and guiding the surgical approach.

The cavernous hemangioma has a clear boundary, and a large cavity is visible inside. The blood flow in the cavity is stagnant, and the red blood cells are separated from the serum. Schwannomas are generally well defined. In this case, the specimen of schwannoma shows that hematoxylin and eosin-stained histologic section of the schwannoma with prominent palisading of the cells, cystic spaces, with S-100 positive and Ki-67 3% positive.

At present, the primary treatment for cavernous hemangioma and schwannomas is surgical resection. For cavernous hemangioma without dysfunction and without affecting appearance, it can be observed regularly without excessive active resection. While schwannomas are different, tumors should be removed early.

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Figure 5. Micrograph of the Schwannoma showed the Antoni B-pattern structures occupying the majority of areas (A, H&E staining, 100×). The expression of S-100 protein and Ki-67 also supports the diagnosis (B, C).

Figure 6. Histopathological examination of the specimen demonstrated cavernous hemangioma with a clear boundary.
Although schwannomas progress slowly, they continue to increase and eventually destroy vision. In this patient, because of the space-occupying lesions in both anterior and posterior part of the orbit, the lateral canthal incision approach was chosen.

Although restricted extraocular motility and proptosis caused by ECMs and schwannomas usually disappear after removal of the lesions. However, the decreased visual acuity and visual field defects may be continuous.

**Conclusion**

Multiple tumors are rare in the same orbit. Heterologous tumors are more unusual at the same time. There are no apparent symptoms in the early stage. When the clinician receives these patients, he should be highly alerted to the initial manifestation of the tumor and should improve the imaging examination for the patient. The tumor oppresses the optic nerve with increasing volume, which may cause vision loss and even blindness. Therefore, even for benign tumors, clinicians and patients should attach great attention to active treatment.

**Disclosure of conflict of interest**

None.

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**References**


