Overview

Cavernous hemangioma (CH) is the most common intraorbital primary benign tumor in adults. The name is a misnomer as it is not actually a tumor but a low flow vascular malformation. Usually CH presents in the second to fourth decades with a female preponderance. These tumors usually present with a painless, progressive axial proptosis. In an uncommon scenario, a CH may lead to restriction of ocular movements and vision loss. Rarely, this lesion may also present with a spontaneous orbital hemorrhage. Characteristically, a CH does not change in size on Valsalva maneuver or with coughing and straining. CH is usually intraconal but can be extracanal also. On magnetic resonance imaging (MRI), the tumor is isointense to muscle on T1 weighted images, hyperintense to muscle on T2 weighted images and shows an inhomogeneous contrast enhancement. It is an angiographically occult tumor. On histopathological examination, a CH has a fibrous capsule with abundant, loosely distributed smooth muscle in the vascular wall and stroma. It consists of thrombosed and septated venous channels arranged in convolutions. Being a vascular malformation, a CH is neither infiltrative nor associated with inflammation of the adjacent tissues but is capable of incorporating adjacent blood vessels and nerves into its capsule as it expands. This non-malignant lesion expands or grows through a series of vascular events. Within the lesion, an intravascular clot forms as vascular stasis and thrombosis occurs. The thrombus formation subsequently leads to endothelial cell proliferation and recanalization of the CH into multiple clefts and vascular channels.

Surgical excision is the treatment of choice for an orbital CH. The common approaches are the transcranial subfrontal approach or a lateral orbitotomy. Both of these approaches require a skin incision and a craniotomy. In recent times, the endonasal endoscopic transethmoid approach is becoming popular for the management of an orbital CH, particularly in patients in whom the CH is located predominantly in the medial part of orbit. The endoscopic approach has certain distinct advantages over the conventional approaches in the form of performance of a scarless procedure and the minimal occurrence of approach-related morbidity.

Surgical Steps

Fig. 1 A 29-year old lady presented with multiple episodes of transient visual blurring and proptosis of the right eye. Examination revealed a mild axial proptosis of the right eye with normal visual acuity of both eyes and intact extraocular movements. Contrast magnetic resonance imaging (MRI) coronal (a) and axial (b) sections show a well-defined lesion in the right orbit causing proptosis (arrow). The mass is intraconal and predominantly located in the medial part of the orbit. It is pushing the optic nerve toward the lateral side.
Fig. 2 Endoscopic view of the right nasal cavity: Adrenaline soaked cotton patties are used to decongest the nasal mucosa.

Fig. 3 After proper decongestion, the middle turbinate (MT), inferior turbinate (IT) and nasal septum (S) are well visualized.

Fig. 4 The inferior turbinate (IT) is lateralized to get the working space in the nasal cavity. The middle turbinate (MT) and the nasal septum (S) are seen.
Fig. 5 The middle turbinate (MT) is medialized to widen the middle meatus. The uncinate process (UP; a thin bone which is a part of ethmoid bone present in the middle meatus. It attaches anteriorly to the posterior edge of the lacrimal bone, and inferiorly to the superior edge of the inferior turbinate [IT]) is seen in the middle meatus.

Fig. 6 Removal of the uncinate process (UP) is performed. The middle turbinate is also visualized.

Fig. 7 After the uncinectomy, the bulla ethmoidalis (BE; bulging of the largest anterior ethmoidal cells) is seen.
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Fig. 8 Opening of the bulla ethmoidalis (BE) is carried out.

Fig. 9 After the bulla ethmoidalis is removed, ground lamella (GL; it is the part of middle turbinate (MT) attached to the lamina papyracea (LP), marking the division between the anterior and posterior ethmoid air cells) is seen. It is made of the second part of middle turbinate. The middle turbinate (MT) is seen medially.

Fig. 10 The ground lamella (GL) is opened to enter into the posterior ethmoidal (PE) air cells.
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Fig. 11 After opening the ground lamella, posterior ethmoidal (PE) air cells are seen. The anterior sphenoidal wall (ASW) is also visualized in proximity to the posterior part of posterior ethmoid air cells. The lamina papyracea (LP) forms the lateral wall of posterior ethmoid air cells. The superior turbinate (ST) forms the medial wall of posterior ethmoidal air cells, and is an important landmark. In the infero-lateral part, maxillary sinus opening (MSO) is seen.

Fig. 12 The superior turbinate (ST) and the anterior sphenoid wall (ASW) are well seen. The posterior ethmoid (PE) air cells and the lamina papyracea (LP) are also seen. The superior turbinate (ST) is lateralized to visualize the sphenoid ostium (SO).

Fig. 13 The anterior sphenoidal wall (ASW) is opened to enter into the sphenoid sinus. The lamina papyracea (LP) and the medially shifted superior turbinate (ST) are also visible.
Fig. 14 The sphenoid sinus (SS) and its mucosa (M) are seen after opening of the anterior sphenoidal wall.

Fig. 15 The sphenoid sinus mucosa (M) is removed with a biopsy forceps. The sphenoid sinus (SS) and lamina papyracea (LP) are also visible.

Fig. 16 After the sphenoidal sinus mucosa is removed, the sphenoid sinus is well seen. The lamina papyracea (LP) is also seen.
**Fig. 17** A close-up view of the inside of the right sphenoid sinus is shown. The lateral optico-carotid recess (LOCR), optic nerve protuberance (ONP), internal carotid artery (ICA) protuberance and intersphenoid septa (ISS) are seen. The lamina papyracea (LP) forms the thin shell of bone between the orbit and the sphenoid sinus.

**Fig. 18** Removal of the thin lamina papyracea (LP) [which can be easily fractured] is being carried out. The periorbita (P) is seen after the lamina papyracea is removed.

**Fig. 19** A close-up view of the periorbita (P) is provided.
Fig. 20  The periorbita (P) is opened by a ball probe.

Fig. 21  The orbital fat (F) prolapses out after the periorbita is opened.

Fig. 22  Dissection of the orbital fat (F) is done to reach the lesion.
Fig. 23 The medial rectus (MR) muscle is seen after the orbital fat (F) dissection.

Fig. 24 After dissecting the orbital fat (F), the cavernous hemangioma (CH) is visualized by lifting up the medial rectus (MR) muscle.

Fig. 25 The cavernous hemangioma (CH) is dissected from all round to make it free from the surrounding structures.
Fig. 26 A gentle bimanual dissection around the cavernous hemangioma (CH) separates it from the surrounding neurovascular structures, bringing it into the nasal cavity. The assistant puts digital pressure over the globe to bring the tumor into the surgical field to facilitate the peritumoral dissection. The displaced medial rectus muscle (MR) is also visible.

Fig. 27 After the tumor is free from the surrounding structures, it is grasped with a biopsy forceps to remove it from the nasal cavity.

Fig. 28 The tumor is being delivered out through the right nostril.
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Fig. 29 Some pieces of surgicel are kept at the operative site for hemostasis.

Fig. 30 A few gelfoam pledges are also kept in the operative area.

Fig. 31 The excised surgical specimen of the cavernous hemangioma is seen.
Complication Avoidance

Ophthalmoparesis and Ptosis

These complications are often transient and recover over a short period of time. These complications can be avoided by a careful, gentle dissection and minimal manipulation of the intraorbital neurovascular structures.

Lid Edema and Conjunctival Hemorrhage

Lid edema and conjunctival hemorrhage both resolve spontaneously over a few days.

Tips and Pearls

- A proper case selection is essential for achieving a good outcome. Only those cases where the CH is located predominantly in the medial part of orbit and shifting the optic nerve laterally, should be addressed with this approach.
- The exposure of the periorbita should be adequate for a proper manipulation of instruments.
- The orbital fat often comes in the surgical field, which can be coagulated and excised to make the surgical field clear.
- The manipulation of extraocular muscles should be gentle to avoid postoperative ophthalmoparesis.
- Great caution should be undertaken while working near the optic nerve to avoid postoperative visual deterioration.

Suggested Readings