Coexistence of Cavernous Hemangioma and Other Vascular Malformations of the Orbit

A Report of Three Cases

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SUMMARY – *Coexistence of orbital cavernous hemangioma and other vascular malformations is unusual and few cases have been reported. We describe the clinical and radiological features of three cases of orbital cavernous hemangiomas associated with other vascular malformations, selected reviewing a series of 181 cases of cavernous hemangiomas. All patients were males (age ranging from 43 to 67 years) without vascular systemic disorders and/or a clinical syndrome. They experienced slow progressive exophthalmos. One of them developed acute pulsatile proptosis (case 2), while another experienced slow progressive diplopia (case 3). In one case vascular lesions were bilateral (case 3) and in two patients two different lesions coexisted in the same orbit (cases 1 and 2). All patients underwent surgical excision, which was partial in two cases. Two patients had cavernous hemangiomas in association with a venous malformation (a varix in case 1 and a lymphangioma in case 2), while in the other ones (case 3) cavernous hemangioma was associated with a low-flow arteriovenous malformation. No patient denied visual impairment postoperatively. Few cases of orbital cavernous hemangiomas coexisting with other vascular malformations have been reported in the literature. This entity seems to be an association of different variants of orbital vascular malformations, presenting with a wide spectrum of clinical forms and probably with the same pathogenesis.*

Introduction

Vascular lesions of the orbit represent 24% of the space-occupying lesions of the orbit, and cavernous hemangioma represents the most common vascular orbital lesion, accounting for 9% of all cases¹. Orbital vascular lesions are generally described as unilateral and intracranial single lesions²⁻⁴. Cavernous hemangiomas may extend to the orbital apex, or have intracranial extension^{5,6}. They seldom present as unilateral multiple lesions^{7,8}, while bilateral lesions are an extremely rare occurrence having been described isolated⁹⁻¹⁴ and associated with systemic diseases like blue rubber bleb nevus syndrome¹⁵, and Maffucci's syndrome^{16,17}. To the best of our knowledge, the coexistence of

cavernous hemangioma and a venous malformation is unusual and has been reported in few cases¹⁸⁻²³. We describe the clinical and imaging features of three unusual cases of orbital cavernous hemangiomas associated with other venous malformations, selected reviewing a series of 181 cases of cavernous hemangiomas.

Case Reports

Case 1

A 63-year-old man had a six-month history of progressive left exophthalmos, with positivity at Valsalva maneuver. A CT scan, previously performed at another Institution (Figure

Figure 1 Case 1. A) A 63-year-old man with left exophthalmos. B) Enhanced CT examination, MPR reconstruction on coronal plane. Two contiguous lesions are visible in the left retrobulbar intraconal space. The larger lesion (white arrow) is an ovoid and well-circumscribed mass, non-homogeneously enhanced; the small lesion (black arrow) is a rounded and well-circumscribed mass.

Figure 2 Case 2. A 74-year-old man with right proptosis.

1), showed two continuous masses in the left retrobulbar intraconal space. The larger one was a well-circumscribed lesion, medially displacing the optic nerve, without invasion. It was an ovoid mass, showing inhomogeneous enhancement. After surgical removal of the larger lesion, histopathological examination showed a cavernous hemangioma. During the surgical intervention, the second lesion was exposed and its macroscopic appearance was consistent with an orbital varix, then it was gently reduced in size by means of partial cauterization.

Figure 3 Coronal Fat Sat mdc (A), coronal FLAIR (B), axial T2-weighted (C) and sagittal SE T1-weighted (D,E) MR images. Two vascular lesions in the right orbit. The largest (small arrow) was a lateral and posterior, rounded and well-defined lesion, displacing the lateral rectus muscle inferiorly, and the superior rectus muscle medially. It appears isointense to the muscle on T1-weighted images and hyperintense on T2-weighted images. The second lesion (large arrow) was multilobulated with an inhomogeneous signal on T2-weighted images and isointense to the muscle on T1-weighted images.

Case 2

A 74-year-old man, who had undergone surgery for removal of a cavernous hemangioma of the right eye 31 years earlier, was admitted to our institution for a rapidly progressive proptosis in the right orbit (Figure 2).

MR examination (1.5T) showed two vascular lesions in the right orbit. The first was a lateral and posterior, rounded and well-defined lesion, displacing the lateral rectus muscle inferiorly, and the superior rectus muscle medially, without invasion.

On MR the lesion appeared isointense to the muscle on T1-weighted images and hyperintense on T2-weighted images. The second lesion was multi-lobulated with an inhomogeneous signal on T2-weighted images. The patient underwent a lateral orbitotomy and both lesions were removed (Figure 3A-E). Histopathological analysis confirmed a cavernous malformation for the larger and posterior lesion, and a venous lymphatic malformation (lymphangioma) for the second lesion. After treatment the patient experienced a complete resolution of the proptosis.

Figure 4 Case 3. A) A 52-year-old man with right proptosis. B) Unenhanced CT examination shows a well-rounded lesion (white arrow) in the lateral portion of the left retrobulbar intraconal space and another (black arrow) not well-defined lesion in the lateral portion of the right orbital apex. C) Axial fat-sat T1-weighted MR image with contrast medium shows significant and inhomogeneous enhancement of both masses.

Case 3

A 52-year-old man had been suffering from intermittent diplopia for five years, and had noticed the onset of a progressive proptosis in the right eye in the last three months (Figure 4A). Clinical examination disclosed a visual acuity of 20/20 bilaterally, limitation of left eye abduction and a slight papilledema on the right eye. CT and MR examinations detected a well-rounded lesion in the lateral portion of the retrobulbar intraconal space, rounded, with homogeneous and intense enhancement after contrast administration (Figure 4B,C). The lesion was removed through a lateral orbitotomy and pathological analysis revealed a cavernous hemangioma. In the right eye, in the lateral portion of the orbital apex, CT showed a not well-defined lesion, with irregular enhance-

ment. This lesion was untreated and on MR-follow-up displayed an inhomogeneous signal on T2-weighted images (Figure 5A-D). It was defined as low-flow vascular malformation.

Discussion

Unilateral and bilateral multiple cavernous hemangiomas have already been described, whereas few literature studies have reported the coexistence of cavernous hemangiomas and vascular malformations¹⁸⁻²³. The incidence of these lesions in our 30-year review is low, accounting for 1.4% among patients with cavernous hemangiomas^{1,2}.

We selected three patients with a cavernous hemangioma coexistent with other orbital vascular lesions such as a venous anomaly, a

Figure 5 Case 3. MR examination on follow-up with axial SE T1-weighted (A), axial TSE T2-weighted (B,C) and axial FLAIR (D) images show the complete excision of the left orbital lesion and the persistence of the right lesions. The lesion appears inhomogeneous on T2-weighted images and isointense to muscular tissue on the T1-weighted image.

venous lymphatic malformation and an arterio-venous malformation (AVM).

As vascular malformations of the orbit present special diagnostic and therapeutic challenges, the hemodynamic characteristics and localization of these lesions play a key role for therapy planning. Vascular malformations of the orbit could be considered vascular hamartomas that derive from the venous system embryologically and have differentiated, in part or whole, from lymphatic or venous vessels. They could best be understood also within the context of their hemodynamics, of which there are four types. Type 1 (no-flow) lesions have essentially little connection to the vascular system and include lymphangiomas or combined venous lymphatic malformations. Type 2 (venous-flow) lesions appear as either distensible lesions with a direct and rich communica-

tion with the venous system or non-distensible anomalies that have minimal communication with the venous system, as varices. Low-flow lesions, type 3, include cavernous hemangiomas, while type 4 are AVM characterized by direct antegrade high-flow through the lesion to the venous side^{24,25}. Thorough clinical and radiological examinations are indispensable for patient counseling.

Cavernous malformations, also known as cavernous hemangiomas, are the most common vascular lesions in adults¹, consisting of enlarged, ectatic (cavernous) venous spaces, surrounded by a capsule of fibrous tissue. They are usually solitary and most often occur in the lateral aspect of the retrobulbar intraconal space. They are rarely intramuscular. Cavernous hemangiomas seldom involve the orbital apex and occasionally may extend intracranially through the

superior orbital fissure and in such cases differential diagnosis could be a challenge²⁴⁻²⁸. Bone remodeling is not uncommon, and intralesional calcification occurs occasionally. Associations with Maffucci's syndrome and blue rubber bleb nevus syndrome have been reported^{15,16}. The main clinical symptoms are slowly progressing, painless, axial exophthalmos, occasionally with mild visual deficits, due to optic nerve compression or extension by stretching; choroidal folds or papilledema are found in some 25% of patients^{29,30}. Imaging characteristics consist of a well-defined, round or oval mass, more frequently intraconal and rarely involving the orbital apex, which may make differential diagnosis from other apical lesions challenging³¹. Calcifications, due to phleboliths, are well-detected by CT. On MR, cavernous hemangioma presents as an iso- to hypointense lesion on T-weighted images, while on T2-weighted series the lesions exhibit a hyperintense homogeneous at times irregular signal. Internal septa are visible within larger lesions. Cavernous malformations demonstrate progressive accumulation of contrast material on late phase dynamic images and delayed images; contrast material does not fill the central part of the lesion until the late venous phase³².

Differential diagnosis of cavernous hemangioma includes orbital varices, also known as orbital venous anomalies. They can appear, on imaging, as well-delineated masses, similar to cavernous hemangioma, but they can be clinically differentiated by the Valsalva maneuver. They result from a congenital weakness in the postcapillary venous wall, leading to the proliferation of venous elements and massive dilatation of the valveless orbital veins³¹. On imaging, orbital venous malformation presents as an intraconal, well-defined, triangular configured mass, tapering toward the apex, with highly intense, homogeneous contrast enhancement^{31,32}. The first case we described here had the peculiar findings of the coexistence of cavernous hemangioma and a varix. Indeed, he underwent a preoperative CT scan, which displayed the two lesions but did not allow differential diagnosis. Thus treatment for this case was a challenge, since initially both lesions were thought to be cavernous hemangioma. Surgery was then performed to remove the two lesions. The larger lesion was removed being a cavernous hemangioma; while only intraoperatively did the second lesion prove to be a varix, which was treated by cauterization to reduce its volume.

Cavernous hemangiomas are generally managed conservatively, and surgical excision is re-

served for those that cause severe proptosis or optic nerve compression. Because of the inaccessibility of the small feeding arteries and the multiple collateral pathways available for recanalization, embolization therapy is not often performed^{31,32}. After surgical removal, orbital cavernous hemangiomas do not usually recur. We recorded one case who presumably had a recurrence (case 2) of the cavernous hemangioma after 31 years. Assuming that cavernous hemangioma was macroscopically removed completely in the first operation, this late recurrence may be explained either by regrowth of a microscopic residual part of the initial lesion or by the new slow growth of a lesion not detectable at the time of first diagnosis because of its size. Indeed, the cases reported in the literature, with incomplete initial resection of the cavernous hemangioma, never experienced recurrence of the lesion over time³¹. Hence, the hypothesis of new growth of the cavernous hemangioma should be considered for this case.

The coexistence of different vascular malformation in the same orbit or in the two orbits has never been widely documented in the literature. In some cases, cavernous hemangiomas can mimic orbital venous lymphatic malformations⁵. In addition, the origin and development of vascular malformations are controversial, most authors claiming that the wide clinical spectrum of these lesions reflects different phenotypes of the same genotype defect at different stages of development³³. This hypothesis is supported by the second and the third cases herein reported of cavernous hemangioma coexisting with a lymphangioma (case 2) and with a low-flow vascular malformation (case 3).

Orbital venous lymphatic malformations, previously known as lymphangiomas, are uncommon and sometimes referred to as no-flow or low-flow vascular malformations. They contain abortive vessels, which spread among normal structures and present as an unencapsulated, primarily thin-walled masses with numerous cystic spaces of different size. They show a tendency to spontaneous hemorrhage, resulting in a sudden onset of proptosis combined with periorbital swelling and reduced eye motility, at times leading to optic nerve compression²⁹. On imaging they present as an infiltrative, multilobulated mass with poor encapsulation, also intra and extraconal, sometimes harboring calcifications seen on CT. MR imaging is the modality of choice for the evaluation of lymphatic malformations because it best depicts the various components.

T1-weighted images best depict lymphatic or proteinaceous fluid, and T1-weighted fat-suppressed images are best for detecting blood or blood products. T2-weighted fat-suppressed images provide improved visibility of components that contain non-hemorrhagic fluid³¹. The use of contrast material does not typically provide significant additional information, but an absence of enhancement is indicative of a lymphatic component. Fluid-fluid levels produced by hemorrhages of various ages within multiple cysts are almost pathognomonic^{32,33}.

AVMs involving the orbit are rare and typically manifest with periorbital swelling, dilated retinal veins and epibulbar vessels, visible or palpable pulsations and an audible bruit. CT and MR with standard and angiographic protocols can help diagnose these lesions³⁴. However, conventional catheter-based angiography typically is required for precise definition and treatment planning^{32,35}.

Our experience indicates that the coexistence of cavernous hemangiomas and other vascular orbital lesions is a fairly frequent event, never previously well-documented. Nevertheless, we should consider that this experience is limited to a small number of patients and could be atypical, not necessarily representing a general behavior. However, we consider

this circumstance is not uncommon and that radiologists should carefully inspect the other orbit for possible concomitant lesions. These cases should be evaluated initially with noninvasive imaging studies, particularly MR with angiographic and dynamic techniques^{31,36}. Conventional angiography is generally warranted only when no structural cause is identified in symptomatic patients or when alteration of blood flow dynamics is suspected and further characterization is necessary.

Our cases were difficult to treat because they required the management of two different orbital lesions at the same time³⁷. Our three patients were surgically treated^{38,39} with removal of the lesion with a macroscopic appearance of cavernous hemangioma. In two cases venous malformations were associated, and in one case there was a low-flow vascular malformation. None all these lesions were removed, without relevant symptoms, so that no further treatment was planned. For these patients with double coexistent vascular lesions, if clinical features are presents, such as visual acuity impairment, diplopia and pulsatile proptosis, the removal of the cavernous hemangioma should be firstly considered in order to reduce the intraorbital pressure and relieve the symptoms.

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