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ORIGINAL ARTICLE

Clinical and Imaging Findings in Multifocal Orbital Vascular Lesions: A Case Series

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ABSTRACT

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INTRODUCTION

A wide variety of vascular lesions may be present in the orbit, displaying a variety of histological features, hemodynamic behavior, imaging characteristics as well as clinical course.¹ Among them, varices and cavernous malformations are often asymptomatic and usually unifocal conditions.^{1,2} We present a series of three patients with multifocal symptomatic orbital varices to highlight the clinical and imaging findings of these lesions.

CASE I

A 58-year-old male presented with a sudden onset of a compensatory head posture (left tilt of the head). A mild pain at the right periocular area was also noted. Systemic and previous ophthalmic history were non-contributory. On clinical ophthalmic examination best corrected visual acuity (BCVA) was 20/20sc in both eyes, the intraocular pressure (IOP) was 12 mmHg and 14 mmHg on the right and left eye (respectively) and slit-lamp biomicroscopy as well as fundoscopy were noncontributory. Hertel exophthalmometry readings were 22 mm (right eye) and 20 mm (left eye). A slight fullness of the right upper eyelid was noted, which was also present on previous photographs (Figure 1). Ocular motility of the right eye was slightly deficient

on supraduction and adduction (Figure 1); however, a clinical neurological examination as well as visual field testing were noncontributory. Forced-duction test revealed mechanical restriction in the supraduction of the right eye. An MRI scan of the orbits was then performed, revealing the presence of multiple cystic vascular lesions in both orbits, along the course of the superior and inferior orbital veins (Figures 2 and 3). The lesions were connected with the stem of the veins and displayed progressive low-grade contrast enhancement (Figures 2 and 3). In some locations, contrast enhancement was most prominent along the walls, compared with the central part of the lesions, suggesting partial central thrombosis. The



FIGURE 1 Patient 1, showing fullness of the right upper eyelid and compensatory head tilt (A). A previous photo also reveals a similar slight asymmetry between the palpebral apertures (B).

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patient was offered the option of the surgical removal of the most distended of the lesions, located along the roof of the right orbit and partly obstructing adduction and supraduction of the right eye but refused and was instead followed conservatively showing progressive improvement in head posture over the following 12 months.

CASE 2

A 60-year-old female presented with progressive painless protrusion to her left lower eyelid over the previous 12 months (Figure 4). Previous systemic and ophthalmic history were noncontributory. BCVA was 20/20 on both eyes and the IOP was 19mmHg and 20mmHg on the right and left eyes, respectively

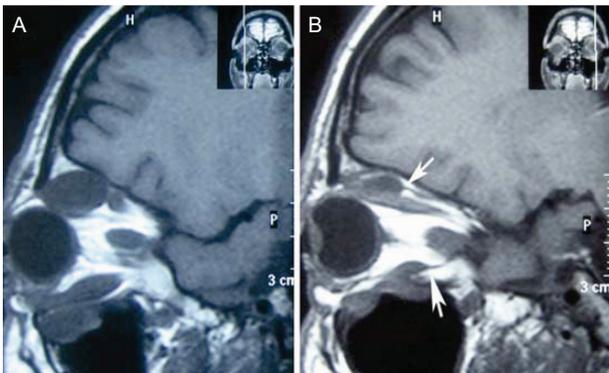


FIGURE 2 T-1 oriented sagittal sections of the right (A) and left (B) orbits of patient 1, showing multiple oval-shaped lesions along the course of the inferior and superior orbital veins. The location of the sections on the coronal plane is shown in insets. The relation of the lesions with the inferior and superior orbital veins is shown with white arrows in B.

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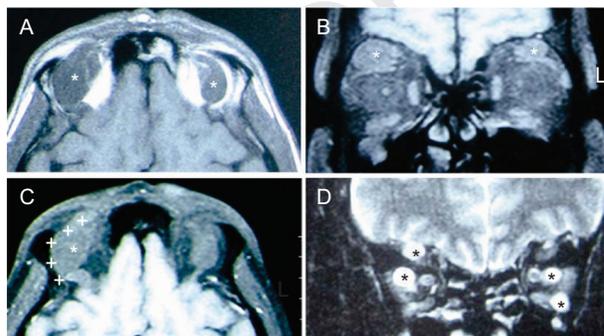


FIGURE 3 T-1 oriented sagittal sections before (A) and after contrast enhancement with fat-saturation (C), as well as T-1 (B) and T-2 (D) oriented coronal sections of patient 1. The saccular distensions of superior orbital veins in both orbits are shown with asterisk. In (C), the core of the lesion displays lower signal enhancement (shown with asterisk) as opposed with the periphery of the lesion which displays higher signal enhancement (shown with "+"), suggesting possible partial thrombosis with associated slow flow.

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(without medications). Slit lamp biomicroscopy and fundoscopy were noncontributory for both eyes whereas ocular motility was full. Hertel exophthalmometry readings were 20mm and 21mm on the right and left eyes, respectively. On valsalva maneuver the protrusion of the left lower eyelid increased considerably. A CT scan of the orbits was performed, revealing the presence of multiple calcified lesions to the left orbit (Figure 4). The lesions were located to the orbital apex, superior-anterior and inferior-anterior parts of the left orbit (Figure 4). The most prominent lesion, located along the inferior-anterior part of the left orbit was surgically excised under general anesthesia. The lesion proved to be an orbital varix associated with a phlebolith (Figure 5). The patient was satisfied with the cosmetic outcome of the procedure and was followed for 12 months without local recurrence.

CASE 3

A 53-year-old female presented with a localized protrusion along the medial aspect of the left lower eyelid, present during the previous 6 months (Figure 6). A mild

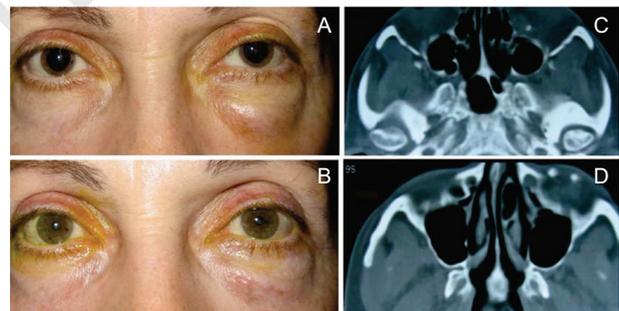


FIGURE 4 Preoperative (A) and postoperative (B) appearances of patient 2. Protrusion of the left lower eyelid was more prominent on Valsalva maneuver (A). Multiple varices of the left orbit with associated phleboliths are shown in transverse CT sections of the superior (B) and middle (D) orbits.

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FIGURE 5 Vascular lesion and associated phlebolith, surgically removed from the left lower eyelid of patient 2 (A). Corresponding CT transverse section of the orbits (B).

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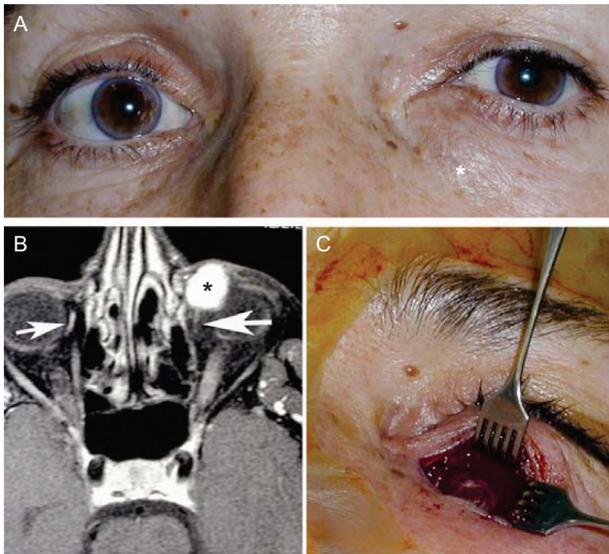


FIGURE 6 Left lower eyelid protrusion in patient 3 (A). T-1 oriented, contrast enhanced and fat-saturated transverse MR section showing a vascular lesion in the inferomedial left orbit (with asterisk) along the course of an orbital vessel (shown with a white arrow in B). A similar enhancing distended vessel is present in the fellow right orbit (also shown with a white arrow in B). Intraoperative view showing a vascular lesion with bluish discoloration (C).

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pain and deformation of the contour of the left lower eyelid were also reported (Figure 6). Previous systemic and ophthalmic history were noncontributory. On examination BCVA was 20/25 and 20/20 on the right and left eyes, respectively whereas the IOP was 15 mmHg on both eyes (without medications). No diplopia was reported and ocular motility was full. Hertel exophthalmometry readings were 19 mm on both eyes. An MRI scan of the orbits was performed, showing the presence of a rounded lesion with significant contrast enhancement along the course of an orbital vein at the infero-medial left orbit (Figure 6). Interestingly, a distended orbital vein at the infero-medial fellow (right) orbit was also present (Figure 6). The lesion of the left orbit was surgically excised under general anesthesia, displaying mixed features of a varix (connections with the inferior orbital vein) and cavernous malformation (fibrous wall and septae). Follow-up showed no recurrence of the lesion or progression of any similar lesion to the right orbit.

DISCUSSION

Orbital cavernous malformations and orbital varices represent a spectrum of vascular conditions characterized hemodynamically by slow blood flow or even stagnant blood (no-flow).³ In the case of cavernous malformations, the anatomic origin has been pro-

posed to be venous or arteriovenous and the lesions are confined by a definite fibrous capsule and divided by septae.^{1,4} On the other hand, varices have communications with the venous orbital system and occasionally become thrombosed due to the low or absent blood flow.^{1,3,5} Clinical findings in the case of cavernous malformations include proptosis, periorbital pain, eyelid swelling, or diplopia whereas in the case of varices proptosis is usually the presenting symptom, often associated with a valsalva maneuver.¹ In this setting, varices may also dramatically present with a sudden onset of painful proptosis (stress proptosis).¹ The age of onset of varices is usually the second or third decade of life, although these lesions probably represent congenital weak sites of the orbital venous system which progressively distend in adult life.⁵ In the first case presented in this report the age of onset was more advanced although the lesions may have been present from younger age, as shown in earlier photos. Interestingly, the presenting symptom in this case was compensatory head tilt, possibly caused by mechanical restriction of supraduction and adduction of the right eye. Furthermore, the varices in MRI scans were multifocal and present in both orbits (which is an unusual finding⁶) and the communications of the lesions with the superior and inferior orbital veins were not extensive (instead, the lesions presented as sacular distensions of respective orbital veins). This might have been related with partial thrombosis of the lesion located at the superior part of the right orbit (as shown in Figure 3).

Although the association of pearly phleboliths with orbital varices has been well described, their presence is often unifocal, occasionally associated with pain or ophthalmoplegia.² On the contrary, in the second case presented in this report the presence of phleboliths at the left orbit was multifocal (each vascular lesion of the left orbit was associated with a calcification) and the presenting symptom was painless protrusion exacerbated with valsalva maneuver, implying that the lesion at the left inferior orbit was not thrombosed, despite the presence of a large phlebolith. The lesion of the third case displayed mixed pathological features of a varix and cavernous malformation although, clinically, the distension with valsalva maneuver was more suggestive of a varix. Interestingly, a distended orbital vein was also detected at the fellow orbit, symmetrically to the site of the affected orbit (infero-medially). This finding supports the possibility that these lesions arise from congenitally weakened or altered vascular structures which may progressively distend to various sizes in adult life.¹

Surgical removal of lesions in patients 2 and 3 was straightforward, with minimal bleeding. Mild intraoperative digital pressure on the globe (as in

blepharoplasty) facilitated prolapse of the lesion in the surgical field which was then detached from its bed with blunt dissection. In the case of patient 1, surgical removal might have been more challenging, since the lesion at the superior right orbit was massive and more definitely connected with the superior orbital vein. The fact that the symptoms associated with this lesion (compensatory head tilt) progressively resolved without surgical intervention implies that conservative management (in the absence of pain or visual field defects) may be a valid option for such lesions.

ACKNOWLEDGMENTS

AQ3 Declaration of Interest:

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