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Case Report

Evaluation of a rare case of orbital venous varix with multiple intracranial venous malformations using prone MRI technique

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ABSTRACT

Orbit is a common site for vascular lesions in all age groups, with venous malformations being the most common type. Orbital varices are a rare subset of venous malformation which is a vascular hamartoma that communicate with normal orbital veins. The mainstay of diagnosis and differentiation from other vascular and neoplastic lesions of orbit is demonstration of change in size of lesion on dynamic maneuvers. In this article, we present a case of left orbital venous varix which showed enlargement on Valsalva maneuver. The patient was evaluated using prone MRI technique to demonstrate increase in the size of lesion. Multiple asymptomatic developmental venous anomalies were detected on brain screening, which is a known association.

Keywords: Orbital varix, Valsalva maneuver, Prone MRI, Developmental venous anomalies

INTRODUCTION

Orbital varices are vascular hamartomas consisting of a plexus of low-pressure, low-flow, distensible thin-walled vessels that communicate with normal orbital veins.[1] Orbital varices are a rare condition, accounting for less than 1.3% of all orbital masses.[2] Most of them are typically unilateral. There is a reported association with intracranial venous anomalies which may or may not directly communicate with the varix.[3] We present a case of left orbital venous varix which showed enlargement on Valsalva maneuver. The patient was evaluated using prone MRI technique to demonstrate increase in the size of lesion. Multiple asymptomatic developmental venous anomalies were detected on brain screening, which is a known association

CASE REPORT

A 42-year-old man presented with chief complaints of swelling and drooping of the left eyelid while bending forward and doing the Valsalva maneuver, which is relieved in the neutral position noticed by him for 11 days. He was evaluated clinically with Valsalva maneuver which demonstrated increase in size of the lesion (Figure 1).

Accordingly, MRI brain and orbit with contrast was planned with acquisition of sequences in supine and prone position to evaluate the change in size of lesion. A well-defined lobulated signal intensity lesion was noted in superior extraconal compartment of left orbit with minimal extension in intraconal compartment near the orbital apex. It was hyperintense on T2/STIR images and showed two tiny T2 hypointense foci within (corresponding to calcific foci on CT screening) suggestive of phleboliths (Figure 2). Dynamic post-contrast study indicates gradual progressive enhancement. The lesion showed definite increase in size in prone position (Figure 3).

On brain screening, multiple developmental venous anomalies (DVAs) with medusa head appearance were noted in bilateral gangliocapsular region and left cerebellar hemisphere (Figure 4). Additionally, there was an enhancing T2/FLAIR hyperintense scalp lesion in subgaleal plane in left parietal region which caused scalloping of underlying bone and communicated with underlying prominent cortical vein through trans-osseous emissary vein. This lesion was suspicious for sinus pericranii (Figure 5).

Figure 1 (A, B): Pre and post Valsalva clinical images demonstrating increase in the size of left orbital lesion with resultant drooping of left eyelid.

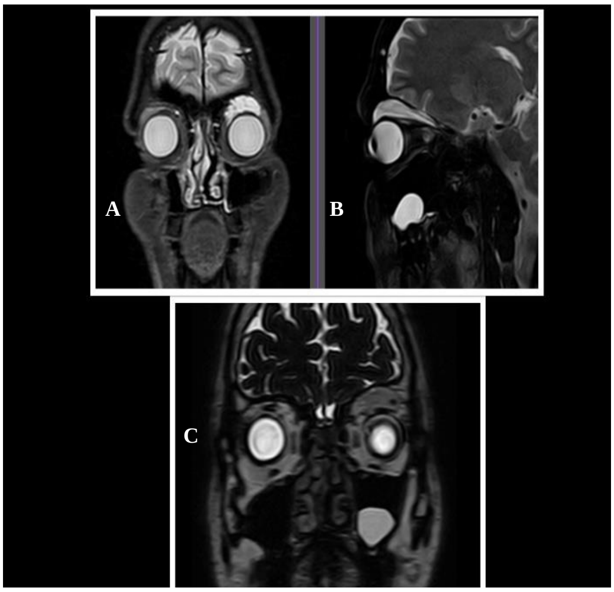

Figure 2 (A-C): Coronal and sagittal stir images showing hyperintense lobulated lesion in superior compartment of left orbit. High resolution 3d-t2wi showing hypointense phlebolith on lateral aspect.

The patient was given options for open surgery versus sclerotherapy for treating the lesion. However, he didn't give consent for either of these, so he was managed conservatively with watchful observation and follow up.

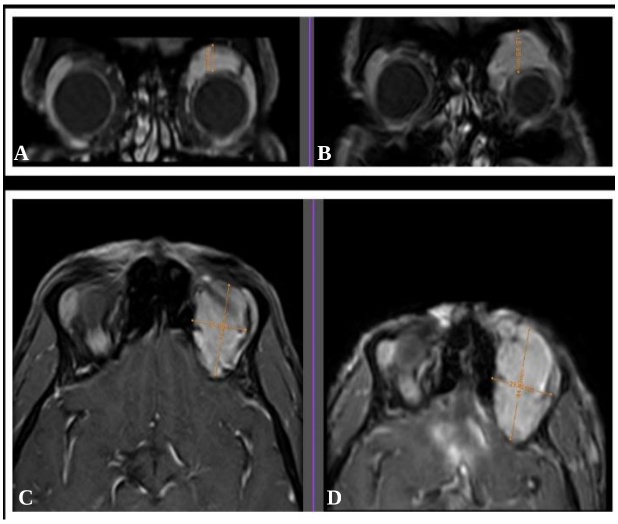

Figure 3 (A-D): Coronal and axial post contrast images in supine (left) and prone (right) positions showing demonstrable increase in size of lesion on prone scan.

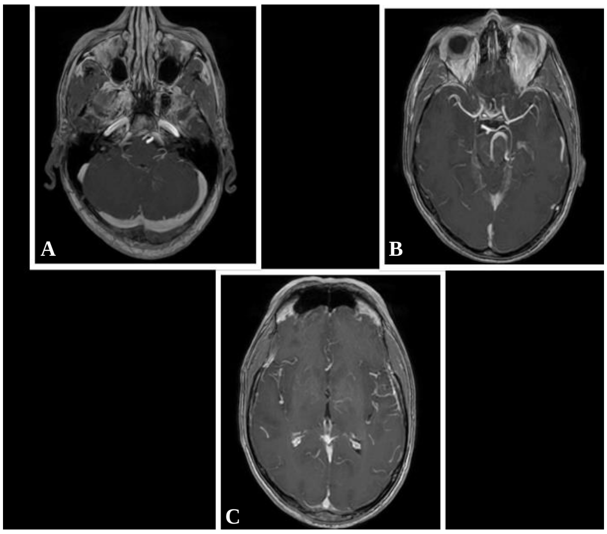

Figure 4 (A-C): images showing enhancing bunch of vessels in left basal ganglia and left cerebellum with a large collector vein in pons suggestive of multiple DVAs.

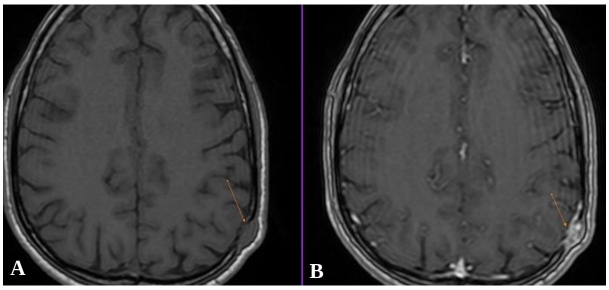

Figure 5 (A, B): Pre and post contrast images showing enhancing scalp lesion in left parietal region with prominent underlying dural vein suggestive of sinus pericranii.

DISCUSSION

Orbital varices are rare vascular hamartomas, typically unilateral and consisting of a plexus of low-pressure, low-flow, distensible thin-walled vessels that communicate with normal orbital veins.[1] They can be divided into primary and secondary types.

Primary orbital varices are idiopathic mostly congenital. Secondary orbital venous varices are those that are acquired due to increased blood flow (as a result of intracranial A-V malformations, carotido-cavernous fistula, dural A-V fistula, etc.) which drain via the orbit.[2,4] These lesions typically present with intermittent diplopia and/or proptosis during episodes of increased venous pressure like straining (Valsalva) or specific positioning such as prone positioning or stooping. There is a reported association with intracranial venous anomalies which may or may not directly communicate with the varix.[3]

Imaging modalities to detect varices include ultrasound, color Doppler imaging, computerized tomography (CT), and MRI including magnetic resonance venography (MRV). During imaging, dynamic maneuvers such as Valsalva, in conjunction with specific positioning, can aid in the visualization of varix.[5] Orbital CT may easily delineate varices and calcified phleboliths, which occur due to thrombus formation. In some instances, orbital varices can erode the surrounding bony structures, leading to orbital expansion and osseous defects of the orbit.[6]

Unenhanced thin-section scanning followed by contrast-enhanced scanning without and with a Valsalva maneuver is the most useful CT technique. Enlargement with a Valsalva maneuver is often associated with orbital varix.

For asymptomatic varices, observation with serial imaging is the usual method of follow-up. For symptomatic varices causing a mass effect, disfigurement, vision loss, optic nerve compression, thrombosis, or hemorrhage, surgical intervention is warranted with a multidisciplinary approach. Generally, orbitotomy with excision of the varices is the treatment of choice. Varices are easier to excise with less invasive modalities if the anterior portion is thrombosed, with identification being more difficult in a supine patient with non-thrombosed varix.[7,8] Recurrence in these cases is usually due to subtotal excision. To avoid this, the surgeon should always aim to resect or clip varix as far back toward the orbital apex as possible.[7,9] Varices can also be embolized for easier identification and to reduce bleeding during surgery. Embolization techniques include the use of micro-coils, as well as the injection of glue, onyx, or cyanoacrylate.[7,10] These techniques are more beneficial in posterior or deeper lesions.

Differential diagnosis includes orbital carotid-cavernous fistula, lymphangioma, AVM, sarcoidosis, pseudotumor and neoplastic masses. Differentiation of vascular and nonvascular lesions is important, because the difference can dictate completely different approaches to further evaluation and management. When vascular lesions are suspected, CT angiography can be helpful for directing further evaluation and management.[11] Vascular orbital lesions tend to have much stronger contrast enhancement than soft-tissue lesions. In addition, attention to morphologic details of the lesions can help differentiate the various types of vascular lesions. Apart from intense contrast enhancement, prominent vessels within or leading to a mass suggest the presence of a vascular lesion.

CONCLUSION

Various imaging modalities serve complimentary role in evaluation of orbital varix. Ultrasound with Doppler can be performed easily with various dynamic maneuvers (e.g. Valsalva maneuver) and in various positions. Orbital CT may easily delineate varices and calcified phleboliths. MRI using prone position which is routinely used for evaluation of tethered cord can be useful to demonstrate dynamic changes in lesion in the settings where prolonged Valsalva maneuver cannot be performed. Screening of brain along with orbit can help detect asymptomatic associated venous malformations and should be incorporated in routine practice.

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