Case Report

Carotid-Cavernous Fistula: Hemodynamic Dysfunction after Shingles? A Case Report and Review Literature

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Case Summary
The patient, a 67-year-old female, presented with esotropia, diplopia, and eye redness, as seen in Figure 1. The diplopia started two months prior to the initial visit while the eye started to crossed in. The patient also complained of severe daily headaches along with ringing in her ears. She believes the symptoms started while she was recovering from shingles on her right side of the forehead. Prior MRI imaging was reported to be unremarkable. The patient was referred to neuro-ophthalmology service with possible diagnosis of thyroid eye disease given the red eye and esotropia. Patient had no history of other medical conditions except shingles in trigeminal distribution that occurred about 4-6 weeks before the onset of her symptoms. Central VA was recorded to be unaffected at 20/20 in both eyes with correction. On sensorimotor exam, there was severe horizontal diplopia at the primary, increasing in left, and right gazes due to bilateral VI nerve palsy. Intraocular pressure testing 22mmHg right eye and 19mmHg left eye, indicating ocular hypertension in the right eye. She had corkscrew blood vessels in both eyes. Optical coherence tomography (OCT) revealed normal retinal nerve fiber layer (RNFL) thickness in both eyes. Visual field and dilated slit lamp examination was unremarkable. Upon closer review of MRI images, significant enlargement of the superior ophthalmic vein (SOV), right more than left was observed. Diagnosis of CCF was made based on typical presentation of corkscrew blood vessels in the eye, bilateral VI nerve palsy, ocular hypertension and enlarged SOV on MORI. A typical bruit was heard by stethoscope over the forehead. New imaging confirmed the arteriovenous malformation (Figure 2). The patient was urgently referred to interventional vascular service. Catheter Angiography showed retrograde drainage in bilateral SOV and bilateral middle cerebral veins. A transcatheter embolization of the left cavernous sinus was performed to repair the type IIb sinus dural AVM with insertion of series of coils. Post procedure angiography revealed no more venous reflux. In one month follow up, the patient’s diplopia improved dramatically. The redness resolved as well as bruit (Figure 1). We are currently monitoring the patient, and expect that the patient will fully recover within the next 6 months.

Figure 1 (Top): Images of frontal photo, right eye, left eye, demonstrating eye redness and strabismus associated with bilateral sixth nerve palsy. (Bottom) Post-operative photos clinically correlated with significant improvement in diplopia.
**Figure 2a:** CT scan showing ophthalmic artery (red arrow) and enhancement of superior ophthalmic vein (blue arrow)

**Figure 2b:** CT scan showing cavernous sinus (blue arrow) concurrently enhancing with cavernous carotid artery (red arrow)

**Discussion**

It is estimated that approximately 76% of all carotid-cavernous fistulas occur as a result of trauma [1,2]. As previously discussed, hemodynamic dysfunction such as hypertension can be a base cause of the development of a carotid-cavernous fistula as well. One such case was reported with clinical presentation of full vision loss and hyphaema [3]. This correlates to the increased rate of CCFs in elderly patients with a history of chronic hypertension and atherosclerosis. Many CC fistula cases resulting from long-term hemodynamic dysfunction as a result of hypertension and atherosclerosis are referred to as spontaneous CC-fistulas or may not be attributed to hypertension or atherosclerosis as the cause of such fistulas. Although most cases are thought to be related to such history of hypertension, atherosclerosis, or trauma, there have been reported cases unrelated to any known etiology. For example, there have been reports of carotid-cavernous fistulas also occurring post-operatively, such as in the cases of trans-sphenoidal operations and carotid thrombectomy [2]. As in our reported patient’s case, one other patient was so far reported with a carotid-cavernous fistula diagnosis with a likely herpes-related etiology. A 73-year-old male had rapid onset of a direct high-flow carotid-cavernous fistula 1 month after a herpes zoster reactivation attack. He presented with diplopia, proptosis, severe frontal headache, and ocular bruit. The patient was treated with endovascular embolization and improved post-operatively [4]. CCFs are still somewhat mysterious and their development has been found to be secondary to a variety of other diseases affecting circulation, from neoplastic growths to aneurysms. Due to the temporal aspect of disease development and absence of significant risk factors for high-flow CCF (direct trauma, etc.), it was presumed that shingles was the likely cause of CCF development in our case. The likely mechanism, considering the patient’s risk factors and history, involved trigeminal nerve gan-
glion HZV reactivation. Due to physical proximity of the ganglion to the artery, arteritis and post-arteritic aneurysm formation could have occurred [4]. Due to such complexity of pathogenesis mechanisms, CCFs remain a complicated disease to diagnose and treat, but with more case reports and literature, such gaps of knowledge can be bridged to better address this patient population [5].

Conclusion
Carotid-cavernous fistulas remain a somewhat uncommon or mysterious occurrence, likely as a result of unclear or unattributable etiologies. Most cases of CCF fistulas occur as a result of direct head trauma or hemodynamic dysfunction related to hypertension or generalized atherosclerosis, especially with chronic conditions seen in the elderly population. In our patient, she developed a severe headache and ophthalmic conditions (ocular hypertension, diplopia, and bilateral scleral injection) 4-6 weeks after reactivation of Trigeminal herpes zoster. CCF was treated with endovascular embolization that resulted in significant improvement. Timely treatment of intraocular hypertension with ocular medications and surgical intervention to repair the CC-fistulas are strongly advised for clinical management of such cases. Should we consider the association of CCF with hemodynamic dysfunction caused by herpes zoster, emphasizes the importance of shingles vaccine in prevention of such complications of herpes zoster re-activation.

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References

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