Case Report

Harlequin Syndrome in Acute Thalamic Hemorrhage

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Harlequin syndrome is a disorder of the autonomic nervous system. It clinically presents as a distinct line of hemifacial sympathetic denervation. We describe a case of Harlequin syndrome with co-existing central first-order Horner syndrome in the setting of a large thalamic hemorrhage with intraventricular extension.

Keywords: Intracerebral hemorrhage—Autonomic nervous system—Harlequin syndrome—Horner syndrome

Background

Harlequin syndrome is a disorder of the autonomic nervous system. It was first described in 1988.[1] It is usually idiopathic, and it follows a benign course.[2] Imaging is needed to outrule a structural cause.[3] It may be associated with other dysautonomias, such as Horner syndrome as seen in this case.[2,4]

Case

A 52-year-old woman was found poorly responsive. She was on aspirin for prior ischemic stroke. On examination, blood pressure was 155/70 mmHg. She was globally aphasic with dense right-sided hemiplegia. A left Horner syndrome with the classic triad of ptosis, miosis and anidrosis was noted. A non-contrast Computed Tomography (CT) brain scan showed a large left thalamic hemorrhage with intraventricular extension. The next day she was hypertensive, distressed, and appeared to be pain. In addition to a left central Horner syndrome, there was intermittent contralateral facial flushing with relative pallor on the ipsilateral side. This finding is called Harlequin syndrome (Fig 1). It resolved with clinical improvement of the intracerebral hemorrhage. Of note, additional imaging including chest radiograph and CT angiogram excluded alternative causes of co-existing Horner syndrome.

Figure 1. *Left Horner syndrome with Harlequin syndrome.*

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Discussion

Harlequin syndrome and Horner syndrome are both disorders of the sympathetic nervous system. They may occur independently or together dependent on the localization of the etiology along the three-order neuron oculosympathetic pathway. The first order or central neuron begins at the posterolateral hypothalamus. The second order or preganglionic neuron exits at the level of C8-T2. Alongside this, sympathetic fibers from the cervicothoracic ganglion are responsible for facial thermoregulatory flushing and sweating. The third order or postganglionic neuron arises from the superior cervical ganglion.[2]

This patient had a central Horner syndrome secondary to external compression of the first order sympathetic neurons arising from the posterolateral hypothalamus. In addition to the classic triad, there was apparent enophthalmus due to sympathetic denervation of superior and inferior tarsal muscles.

This patient also had Harlequin syndrome; a distinct line of hemifacial sympathetic denervation. At times of sympathetic activation the denervated side remained pale and dry in stark contrast to the innervated side, which was moist and red. It is worth noting that there is no general consensus in nomenclature; "Harlequin sign" and "Harlequin syndrome" are both used interchangeably in the literature.

Conclusion

Harlequin syndrome in association with a central first-order Horner syndrome is a novel observation in the setting of an acute thalamic hemorrhage.[5]

References

1. Lance JW, Drummond PD, Gandevia SC, et al. Harlequin syndrome: the sudden onset of unilateral flushing and sweating. J Neurol Neurosurg Psychiatry 1988;51(5):635-642.
2. Brazi P, Masdeu J, Biller J. Localizations in clinical neurology. 6th ed. Philadelphia: Lippincott Williams & Wilkins; 2011.
3. Drexler I, Traenka C, von Hessling A, et al. Internal carotid artery dissection and asymmetrical facial flushing: the Harlequin sign. Stroke 2014;45:e78-e80.
4. Wasner G, Maag R, Ludwig J, et al. Harlequin syndrome – one face of many etiologies. Nat Clin Pract Neurol 2005;1:54-59.
5. Blondin NA, Schindler JL. Teaching neuroimages: first-order Horner syndrome due to ipsilateral thalamic hemorrhage. Neurology 2011;76:14.