Teaching Neuro*Images*: Harlequin syndrome caused by lesion of sympathetic regulatory neurons

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A 48-year-old woman presented with a 6-year history of recurrent episodes of exertional asymmetric flushing of the face and head. After exercise, she observed a distinct line of demarcation between the left half of her face, which was red, and the right half, which retained its normal color (figure, A). Decreased right facial temperature and sweating were also noted. These episodes resolved after 1 hour of rest. She had no ptosis or myosis. She had mild trauma to the right neck 2 years prior to this complaint.

MRI of head, neck, and cervicobrachial plexus were normal. CT angiogram of the right internal carotid artery showed a small segmented stenosis (figure, B and C). Loss of flushing on one side of the face indicates an ipsilateral lesion of sympathetic neurons innervating the face. The absence of Horner syndrome indicates intact oculosympathetic fibers.[1,2] This rare and clinically striking syndrome may result from occult carotid dissection.[3]

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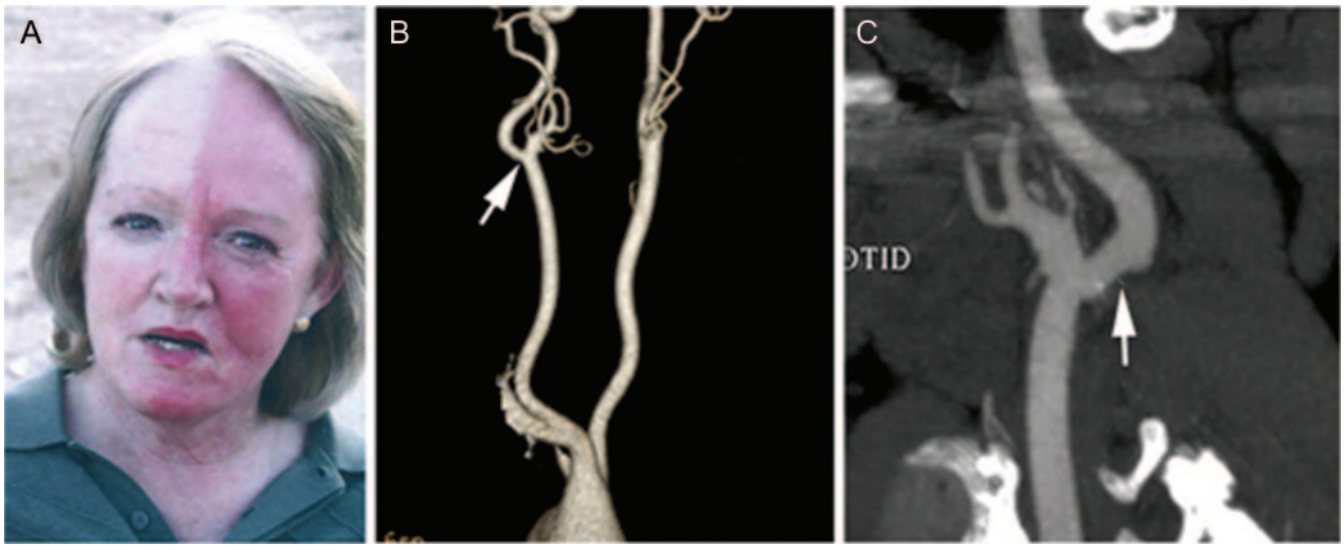
Figure Clinical and radiologic features of harlequin syndrome



(A) The patient after exertion showing loss of right-sided flushing and sweating of the face and head. The left side showed normal flushing. (B) Three-dimensional and (C) 2-dimensional CT angiogram of the carotid artery showing stenosis at the origin of the right internal carotid artery.

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