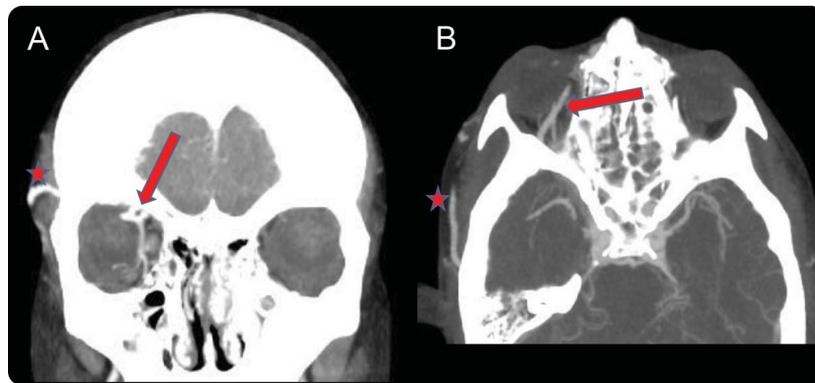


Teaching NeuroImages: Carotid cavernous fistula in a patient with Ehlers-Danlos syndrome

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Figure Head CT angiography



Asymmetric prominence of right superior ophthalmic vein (A, arrow) within the orbit and right frontal orbital scalp vein (B, star).

A 25-year-old woman with Ehlers-Danlos syndrome type IV and history of spontaneous right petrous internal carotid artery dissection presents with severe headache, right eye pain with extraocular movements, proptosis, and chemosis of the right eye. She reported hearing her heartbeat in her right ear. CT angiogram (figure) revealed an engorged right ophthalmic vein. Subsequent digital subtraction angiography confirmed a carotid cavernous fistula along a previously dissected carotid artery.

Unilateral pulsatile tinnitus and eye pain in any patient, especially those with collagen vascular disease, should raise suspicion for a carotid cavernous fistula. Unilateral enlargement of the

superior ophthalmic vein is a distinctive radiographic sign.¹

AUTHOR CONTRIBUTIONS

John W. Liang: acquisition of data, clinical data review, literature review, final manuscript writing. Deborah Horowitz: final manuscript writing.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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