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Non-traumatic carotid dissection and stroke associated with anti-phospholipid antibody syndrome: Report of a case and review of the literature

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Young adults with stroke frequently do not have any of the traditional risk factors associated with stroke, prompting a search for other mechanical and hypercoagulable causes. The authors report a young man presenting with stroke and subsequently diagnosed with a carotid dissection. Recurrent strokes while on heparin prompted a search for a second etiology and the patient was found to have antiphospholipid antibody syndrome. Although these conditions may be coincidental, we propose that their interaction was significant in this patient's presentation. Other reports of this association will also be reviewed.

Key words: Antiphospholipid antibody syndrome, carotid dissection, stroke

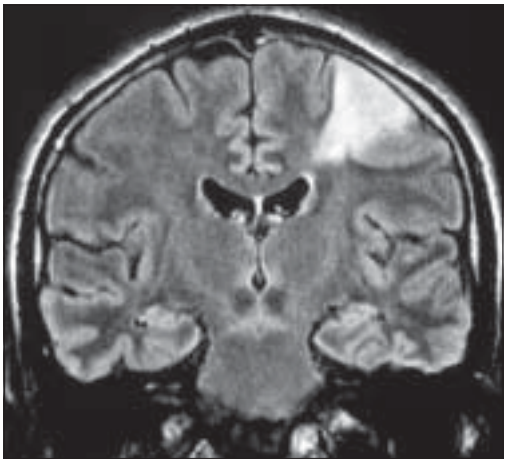

Figure 1: MRI coronal flair image of a wedge-shaped left hemispheric infarction consistent with an embolic source

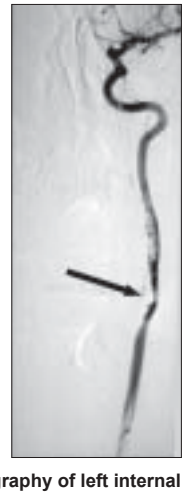

Figure 2: Contrast angiography of left internal carotid artery demonstrating a dissection (arrow)

A 41-year-old right-handed man presented to an emergency department with the acute onset of a left peri-orbital headache and halting speech. His evaluation included a reportedly normal head CT and a urine toxicology screen positive for tetrahydrocanabinol (THC). He was discharged with a diagnosis of THC intoxication.

Eight days later we saw him in our Neurology Clinic. There had been no change in his symptoms. His past medical history was negative and he denied any recent trauma. He admitted to occasional marijuana and alcohol use and smoked two packs of cigarettes per day. On neurological examination he had left eye ptosis, meiosis and a disfluent aphasia. His original CT scan was reviewed and showed evidence of a left subcortical hypodensity. An urgent MRI demonstrated a left-sided stroke [Figure 1] and suggested a left internal carotid artery dissection (CAD) which was confirmed on contrast angiography [Figure 2].

The patient was admitted to the hospital and treated with intravenous heparin. His baseline partial thromboplastin time (PTT) was elevated at 110. His physicians believed that this value had been obtained on heparin and lowered the infusion rate. Four days after admission he woke with global aphasia and right hemiparesis. An emergent head CT demonstrated an

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extension of his stroke. His heparin levels suggested that his anticoagulation with heparin was inadequate and that his PTT was elevated due to antiphospholipid antibodies. Coagulation studies demonstrated the repeated presence of anticardiolipid antibodies, beta-2 glycoprotein antibody and positive mixing studies and dilute Russell Viper Venom Test on repeated testing 12 weeks apart, thus meeting full international criteria for antiphospholipid antibody syndrome (APLAS).[1]

This patient presented with classical signs and symptoms of CAD. Of interest is the coincident hypercoagulable state. While APLAS is a known cause of stroke in young adults it is unlikely that further diagnostic testing would have been done were it not for his clinical course.[2]

Our knowledge of the risk factors for CAD is drawn from symptomatic patients. It is possible that there are also asymptomatic dissections. A two-hit hypothesis posits that while the majority of patients with isolated CAD will remain asymptomatic those patients with another vascular risk factor will be at a significantly increased risk for an ischemic presentation. Sibilia *et al*. reported two cases of stroke associated with CAD and APLAS.[3] He also reviewed three further cases, as well as four patients with antiphospholipid antibodies who did not meet the full criteria for APLAS. Symptomatic CAD has also been associated with Factor V Leiden, MTHFR genotype and elevated homocysteine.[4,5] Further support for the two-hit hypothesis comes from studies of stroke in patients with patent foramen ovale. These studies demonstrate an overrepresentation of hypercoagulable disorders, suggesting that the associated coagulopathy is important in the etiopathogenesis.[6]

Alternatively, APLAS and CAD may be causally linked. Antiphospholipid antibody syndrome induces endothelial damage that may predispose affected arteries toward spontaneous dissections. Conversely, arterial injury from dissection may expose the immune system to hidden antigens which could precipitate a hypercoagulable state. As antiphospholipid antibodies are present in 3-6% of normal individuals these findings may be entirely coincidental.[7] Further epidemiological studies will be needed to determine the relevance of this association.

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