The role of high resolution-vessel wall imaging in the diagnosis of moyamoya disease presenting atypically.

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Introduction: Moyamoya disease (MMD) is a progressive steno-occlusive disease of the terminal portions of internal carotid arteries (ICAs), and proximal cerebral arteries, which leads to collaterals formation. It is typically bilateral, but unilateral MMD can occur. Early stage MMD can present with the absence of the characteristic moyamoya collaterals, making a reliable diagnosis challenging. We explored the utility of high-resolution vessel-wall MRI (HR-VWI) in the diagnosis of large-vessel vasculopathy. Case report: A 45-year-old right-handed man with history of recurrent left hemispheric cerebral infarction taking aspirin, rivaroxaban and atorvastatin, presented to the ED with headache, transient fluent aphasia, and right hemiparesis. PMH revealed factor V Leiden and prothrombin G20210a heterozygosity. Neurological exam was positive for dysgraphia, dyscalculia, finger agnosia, apraxia, and right facial paralysis. Brain MRI showed no acute infarction. MRA showed terminal left ICA, proximal left ACA/MCA stenosis. Differential diagnosis included vasculitis, MMD, and accelerated atherosclerosis. His lipid profile, hemoglobin electrophoresis, and autoimmune studies were normal. Catheter angiography showed unilateral Suzuki stage 2. Finally, HR-VWI revealed a vasculopathy with inward (negative) remodeling and no enhancement on the left supraclinoid ICA, proximal A1, and M1 segments (Fig.1).