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 (1). Early stage MMD can present with the absence of the characteristic Moyamoya collaterals, making a reliable diagnosis challenging (2). We explored the utility of high-resolution vessel-wall MRI (HR-VWI) in the diagnosis of large-vessel vasculopathy.

CASE

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. His vital signs were stable. Neurological exam was positive for left to right disorientation, 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 (Fig. 1). Finally, HR-VWI revealed a vasculopathy with inward (negative) remodeling and no enhancement on the left supraclinoid ICA, proximal A1, and M1 segments (Fig. 2).

DISCUSSION

The diagnosis of MMD can be challenging in patients with unilateral involvement, and no obvious network of collaterals. In fact, some studies report that unilateral MMD can be fairly common considering that it tends to progress to bilateral disease (3) . These findings are common in early-stage MMD, making it difficult to differentiate MMD from other mechanisms of steno-occlusive disease like atherosclerosis or vasculitis. However, this distinction is imperative since MMD benefit from surgical approaches, while the others are managed medically (4). Recently, HR-VWI has emerged as a tool to differentiate the negative remodeling with mild or no concentric enhancement of MMD (highly sensitive and accurate) (5) from the positive remodeling with eccentric heterogeneous enhancement of atherosclerosis and the smooth concentric homogeneous enhancement of vasculitis (6). In conclusion, HR-VWI may be useful for the diagnosis MMD especially on early stages, where unilateral disease is common.

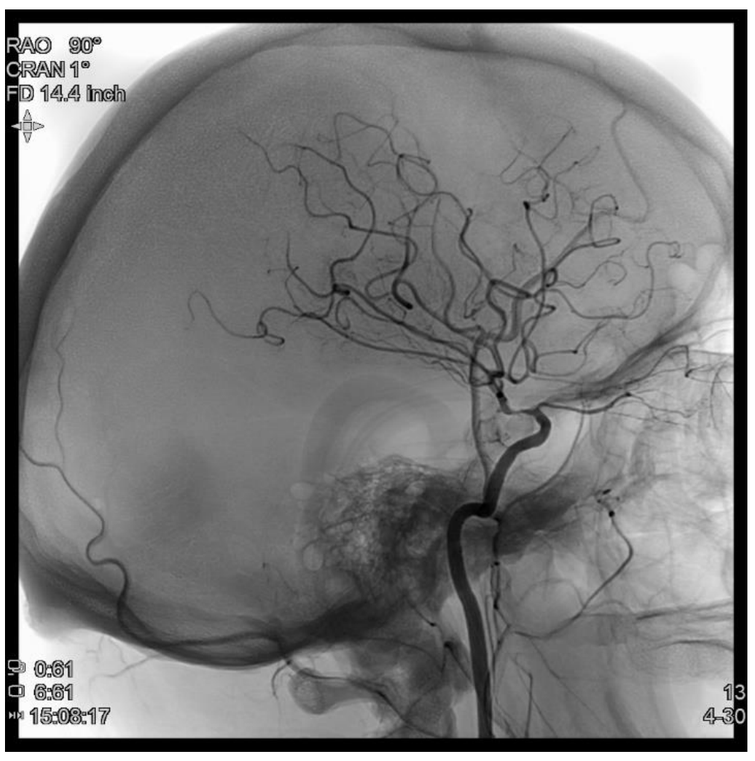

Figure 1. Catheter cerebral angiogram. Left ICA injection exhibiting unilateral Suzuki stage 2 lesion.

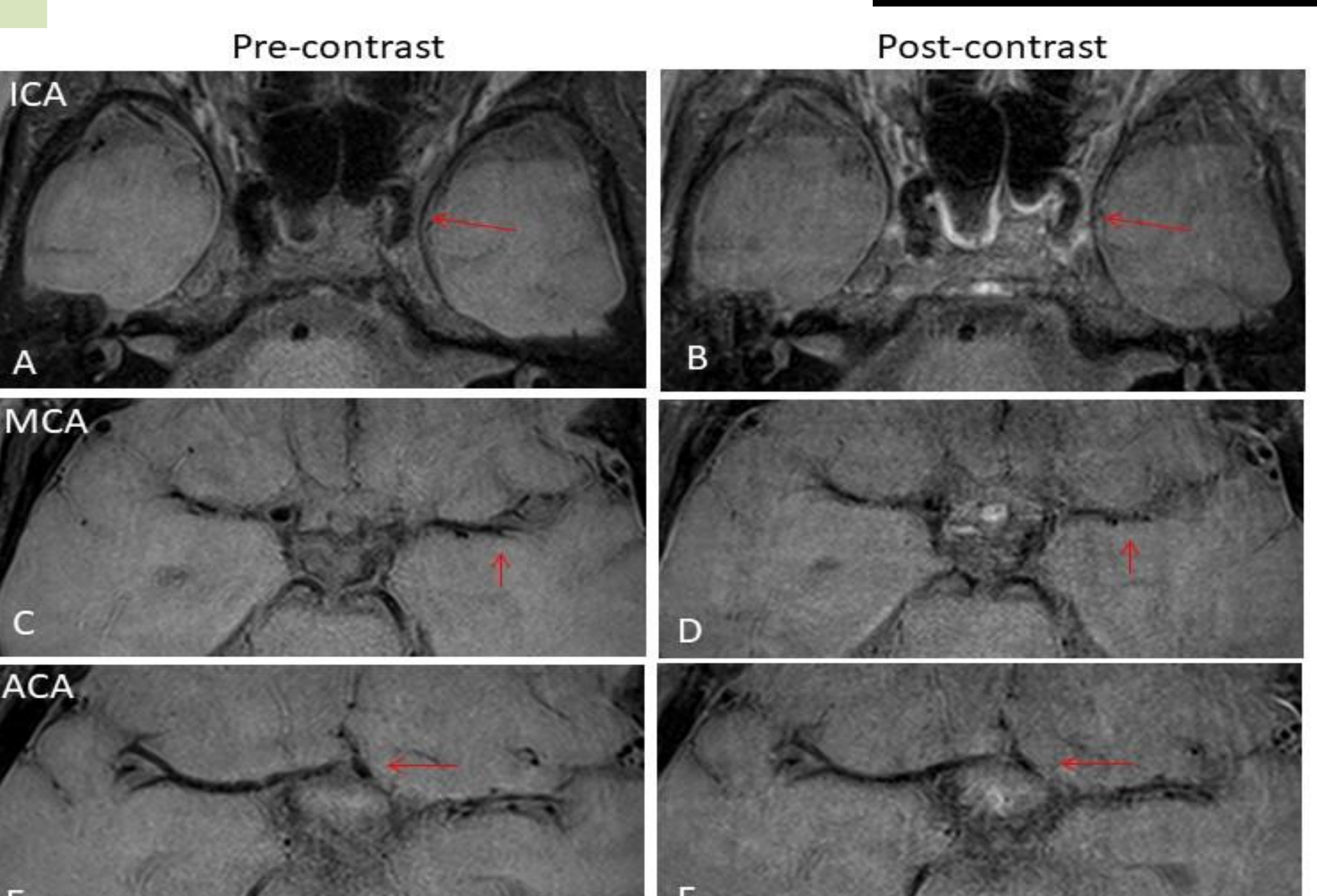

Figure 2. High-resolution vessel wall imaging PANELS A, C, and E show stenotic lesions on left ICA, MCA, ACA. PANELS B, D, and F show the lack of contrast enhancement on the same lesions

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