High-resolution Vessel Wall MRI: A New Armamentarium for Diagnosis of Primary Angiitis of CNS

Vessel wall magnetic resonance imaging (MRI) has gained considerable acceptance and popularity in the diagnosis of various intracranial pathologies.[1] Most commonly used sequences in vessel wall MRI are three-dimensional (3D) fast spin-echo sequences with variable flip angle refocusing pulses. These provide reasonably good spatial resolution to image the circle of Willis with a scan duration ranging from 5 to 10 min.[2] Multiple modifications of the technique have been described in the literature. From its initial use in the diagnosis of carotid plaque morphology, vessel wall imaging has now widened its clinical horizon in diagnosis and prognostication of various intracranial pathologies, such as atherosclerotic disease, moyamoya, central nervous system (CNS) vasculitis, and dissection.[2]

Primary angiitis of CNS (PACNS) is a poorly understood rare disorder with nonspecific clinical presentation.[3] It often presents as a diagnostic challenge to the treating physician.

A diagnostic criterion for the diagnosis of PACNS was proposed by Calabrese and Mallek in 1988. These were later modified by Birnbaum *et al*.[4] to provide "probable" and "definite" diagnoses.

The present gold standard for imaging in PACNS is digital subtraction angiography with a reported sensitivity varying from between 40 and 90% due to different subtypes and patterns of vascular involvement in PACNS.[5] The Digital subtraction angiography (DSA) has low specificity as the main information provided is based on changes in vessel contours without any insight onto the underlying pathological process Thus many diseases, such as RCVS, atherosclerosis, or CNS vasculitis can have similar imaging findings on DSA. Brain biopsy with adequate sampling can establish a definite diagnosis of PACNS. However, the sensitivity of brain biopsy is also low ranging from 54–83% again reflecting the different pathological spectrum of the disease.[6,7] Thus, patients with small vessel patterns of disease are frequently DSA negative and those with medium vessel patterns of the disease may have an inconclusive biopsy.

High-resolution vessel wall MRI is a promising tool in the diagnosis of PACNS. It provides a direct morphological assessment of the vessel wall in addition to the contour abnormalities. Few important considerations in vessel wall MRI are pertinent for adequate interpretation and clinical use, such as contrast to noise ratio, spatial resolution, magnet strength, use of appropriate pulse sequences, and utilization of two-dimensional vs 3D sequences.[2]

With the current resolution, vessel wall MRI is capable of detecting PACNS with medium vessel wall subtype involvement. But with ongoing research, it may be possible to image changes in distal small vessels soon. Sequential imaging and monitoring of factors, such as degree of enhancement and remodeling, it may be possible to evaluate the treatment response after therapy or prognosticate over the clinical course.[7]

On vessel wall MRI, PACNS shows the presence of smooth, concentric wall thickening with strong enhancement. Intracranial atherosclerotic disease (ICAD) shows eccentric, irregular, and short-segment wall thickening with mild enhancement depending on the plaque morphology. Reversible cerebral vasoconstriction syndrome (RCVS) can have a similar angiographic pattern; however, vessel wall MRI can demonstrate a lack of enhancement in these cases which is usually present in PACNS.[1,2,7]

In this issue of Annals of Indian Academy of Neurology, Sundaram *et al*.[8] have demonstrated the clinical utility of vessel wall MRI in the diagnosis of PACNS. They have demonstrated patterns of concentric wall thickening in 12 (60%), diffuse enhancement in 17 (85%), and negative remodeling in 11 (55%) of their study population. Although the study population in the current study is small, however, it strongly demonstrates the clinical usefulness of vessel wall MRI as a noninvasive diagnostic tool for PACNS. They have also demonstrated the negative remodeling in more than half of their study population which can be a pointer to differentiate from ICAD.

There is some overlap in the pattern of thickening, enhancement, and remodeling in different intracranial vascular pathologies as assessed with vessel wall MRI, still, it appears to be a promising diagnostic tool in the diagnosis of PACNS.

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