State of the Art Imaging in Autoimmune Cerebral Vasculitis

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Introduction

Autoimmune cerebral vasculitis can be one of the most devastating diseases as well as one of the most difficult diseases to diagnose. While histopathology is required for the most definitive diagnosis, imaging can provide useful clues to narrow the differential diagnosis and hasten appropriate therapy.

Classification of cerebral vasculitis

A common classification of cerebral vasculitis uses the size of the affected arteries and the etiology of the wall inflammation, but this classification scheme was established for systemic disease rather than cerebral disease \cite{1,2,3,4}. Another way to classify cerebral vasculitis has been proposed by Kuker \cite{4} dividing it into large vessel vasculitis (LVV), medium vessel vasculitis (MVV), and small vessel vasculitis (SVV). The LVV is defined as affecting the ICA, the M1 and A1 segments, the intracranial vertebral artery, basilar artery and the P1 segment. MVV affects the arteriographically demonstrable arteries distal to the M1, P1, and A1 segments. SVV affects the small arteries beyond the spatial resolution of most conventional cerebral angiography. Some etiologies of vasculitis affect a combination of different sized vessels. In this presentation, we will focus on autoimmune vasculitis, for example, systemic lupus erythematosis (SLE), Sjogrens, and rheumatoid arthritis, which mainly affect small vessels.

Imaging methods

Magnetic resonance imaging (MRI) is the method of choice for imaging patients with suspected cerebral vasculitis. The routine MRI protocol for this indication should include pre and post contrast T1-weighted images, T2*, T2-weighted, fluid attenuated inversion recovery (FLAIR) images, as well as diffusion weighted images (DWI). In addition, MR angiography (MRA) and CT angiography (CTA) of the intracranial vessels can be helpful in LVV. Other MRI techniques that might be helpful include MR perfusion, especially if larger vessels are involved, MR spectroscopy (MRS), a technique measuring concentrations of different brain metabolites, and diffusion tensor imaging (DTI), a technique evaluating water indices such as apparent diffusion coefficient and fractional anisotropy.
Conventional cerebral angiography is required to reliably demonstrate lesions in medium sized and small vessels. However, inflammatory lesions in very small cerebral arteries remain below the resolution of most conventional angiography. The difficulty in identifying cerebral vasculitis, and differentiating it from other causes of arterial irregularity and stenosis, explains the numerous false negative cerebral angiograms often seen in this patient population.

**Imaging findings**

MRI findings in autoimmune cerebral vasculitis are, in many cases, nonspecific and include multiple ischemic brain lesions of different ages present in different vascular territories, intra-axial or subarachnoid hemorrhage, and areas of diffuse or discrete pathologic contrast enhancement [5]. MRA and CTA might demonstrate focal stenosis and irregularities of the larger arteries, however MRA and CTA findings in vasculitis are often normal or non-specific since the evaluation of the smaller intracranial vessels is not reliable on these modalities. Conventional angiography can help in some cases, but the final diagnosis is most often made by brain biopsy.

Several MRS studies have demonstrated abnormal brain metabolite concentrations with decreased N-acetylaspartate (NAA), a neuronal marker, and elevated choline, suggestive of increased metabolic turnover, in patients with neuropsychiatric SLE (NP-SLE) [6,7]. In a recent study utilizing DTI, abnormal water movement was seen in normal appearing white matter in NP-SLE patients and, more recently, also in SLE patients without neurological symptoms [8,9]. Increased whole brain diffusivity has also been demonstrated in both SLE and NP-SLE patients in two studies [10,11]. These findings suggest that MRS and DTI are promising tools in the evaluation of patients with vasculitis.

This presentation will focus on conventional MRI techniques and imaging findings in cerebral vasculitis. However, more advanced MR imaging methods, such MRS, magnetization transfer, and DTI will also be discussed.

**References**


