

Isolated middle cerebral artery stenosis caused by syphilitic vasculitis

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Handling Editor:

Somesh Gupta

Received: 8 September 2022
Accepted: 16 November 2022
Published: 2 December 2022

Cite this:

Zhang P et al. (2023)
Sexual Health, **20**(1), 80–82.
doi:[10.1071/SH22147](https://doi.org/10.1071/SH22147)

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ABSTRACT

Background. Syphilitic vasculitis (SV) is rare and difficult to be diagnosed. High-resolution vessel wall magnetic resonance imaging (HR-VWI) might be valuable in the diagnosis by differentiating SV from other vessel wall pathologies. **Methods.** We report a rare case with severe isolated middle cerebral artery (MCA) stenosis and cerebral infarction, which was evaluated by serial HR-VWI. **Results.** A 46-year-old woman presented with an acute infarct in the left basal ganglia and severe isolated stenosis in left middle cerebral artery (MCA). Cerebrospinal fluid (CSF) analysis was performed and the findings were consistent with neurosyphilis diagnosis. The MCA stenosis was also suspected to be the result of SV. HR-VWI revealed the evidence of concentric thickening and circular uniform enhancement of the arterial wall. After two cycles of anti-syphilis treatment, subsequent HR-VWI showed that the MCA stenosis was reduced significantly except slight enhancement of the arterial wall, which confirmed that the SV caused the isolated MCA stenosis. **Conclusions.** HR-VWI could be an important diagnostic tool for isolated middle cerebral artery stenosis caused by SV, which could serve as a biomarker for assessing the response to anti-syphilis treatment in the future.

Keywords: cerebral infarction, cerebrospinal fluid, diagnosis, magnetic resonance imaging, middle cerebral artery, penicillin G, syphilis, vasculitis.

Introduction

Syphilitic vasculitis (SV) is an uncommon cause of cerebral infarction and intracranial vascular stenosis. For these patients, the general treatment for cerebral infarction may be ineffective. Therefore, the identification of SV during the early stages is essential. The current diagnosis of neurosyphilis are mainly relied on cerebrospinal fluid tests, such as the rapid plasma reagin (RPR), the venereal disease research laboratory (VDRL) and the toluidine red unheated serum test (TRUST).¹ However, cerebrospinal fluid tests are not commonly done with patients with cerebral infarction. When patients present with isolated intracranial vascular stenosis, it is difficult to differentiate SV from other vascular pathologies. High-resolution vessel wall magnetic resonance imaging (HR-VWI) has been successfully employed in the characterisation of intracranial vessel wall pathology.² HR-VWI does not only allow wall and lumen imaging, but also provides important information in the structure of vessel wall to hint the causes of vascular stenosis. We reported a case with cerebral infarction and isolated middle cerebral artery stenosis caused by SV and followed up the HR-VWI changes after anti-syphilis treatment.

Case report

A 46-year-old woman was admitted to our neurovascular centre due to weakness in her right limbs, which began while doing routine housework 6 days earlier. Three days earlier, her speech became slurred and prompted her to visit her doctor. No significant atherosclerotic risk factors were found, but physical examination showed that the corner

of her mouth tilted to the left side and the tongue deviated to the right side. The muscle strength of her right limbs was grade 4/5, and the right Babinski sign was positive. Magnetic resonance imaging (MRI) scan showed cerebral infarction in the left basal ganglia, and the brain magnetic resonance angiography (MRA) scan revealed severe stenosis of left MCA M1 segment (Fig. 1). A diagnosis of cerebral infarction and left MCA stenosis was made. Admission serum treponema pallidum particle agglutination (TPPA) and toluidine red unheated serum test (TRUST, 1:2) were both positive. Cerebrospinal fluid (CSF) analysis disclosed elevation of the leukocyte count ($18 \times 10^6/L$) and protein content (1162 mg/L). CSF Venereal Disease Research Laboratory (VDRL, 1:2) was positive. Bacteria and other microorganisms including fungi were negative. To identify the cause for isolated left MCA stenosis, HR-VWI was performed, which demonstrated that the concentric arterial wall was thickened and showed evidence of diffuse gadolinium enhancement of left MCA M1 segment (Fig. 1). This suggested that inflammatory changes may be involved. In the context of the laboratory and neuroradiological findings, a diagnosis was made of active neurosyphilis with vascular involvement, as well as left MCA stenosis and cerebral infarction caused by SV. The patient was treated with clopidogrel (75 mg/day) and aspirin (100 mg/day) for 3 weeks, and intravenous penicillin G (4 MU/4 h for 14 days) at 3-month intervals. After two cycles of penicillin treatment, CSF VDRL became negative, with the decline of leukocyte count ($8 \times 10^6/L$) and protein content (509 mg/L) in CSF. HR-VWI performed at 6 months after the second cycle of penicillin treatment revealed only slight stenosis of left MCA M1 segment and slight enhancement of the wall of the stenosed segment, although it remained mildly thickened (Fig. 2). She was left with slight right limbs weakness that did not affect her life and work.

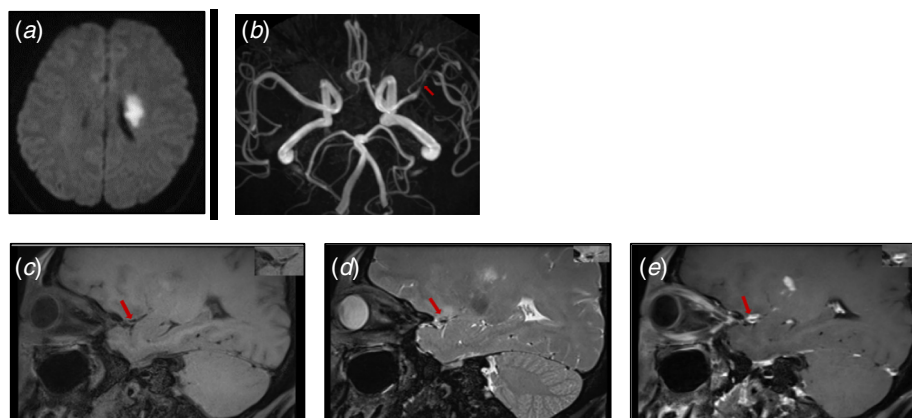


Fig. 1. Representative MRI and MRA images before anti-syphilis treatment. (a) Diffusion-weighted image shows hyper-intense signals of the left basal ganglia. (b) MRA shows severe stenosis in the left MCA M1 segment. (c, d) HR-VWI shows concentric arterial wall thickening in the left MCA M1 segment. (e) HR-VWI demonstrates circular uniform enhancement in the left MCA M1 segment.

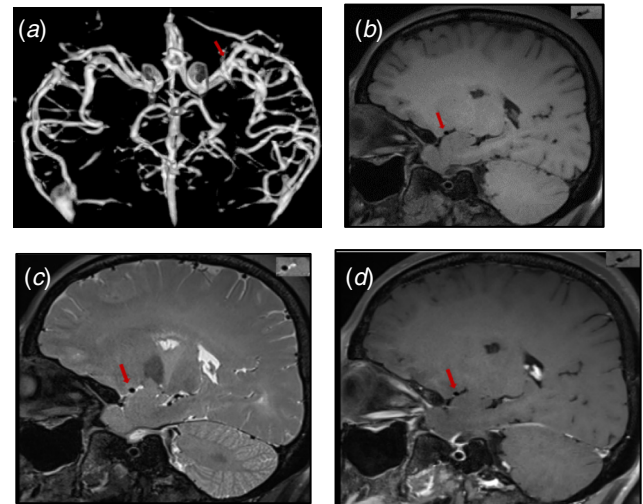


Fig. 2. Representative MRA and HR-VWI images 6 months after the second cycle of penicillin treatment. (a) MRA shows only slight stenosis in the left MCA M1 segment. (b, c) HR-VWI shows mild arterial wall thickening in the left MCA M1 segment. (d) HR-VWI demonstrates only slight enhancement in the left MCA M1 segment.

Discussion

SV can occur at any stage of syphilis and can limit a patient's physical mobility. SV is often misdiagnosed because of non-specific symptoms and diverse forms, especially when there is lack of a clear medical history. SV induces vessel intimal proliferation. Depending on the calibre of the vessels involved, SV can manifest in two forms: (1) Nissl–Alzheimer type, affecting small-calibre vessels; and (2) Heubner type, affecting medium-sized vessels.³ MRI is considered as the most sensitive imaging method in detecting the changes of cerebral vasculitis, approaching 100%, but with a lower

specificity. HR-VWI may help identify which form of SV is involved. In our case, the left MCA showed concentric wall thickening with circular uniform enhancement and was considered to have the Heubner-type involving MCA M1 segment. Because the left basal ganglia receives its blood supply from the lenticulostriate arteries that originate from the left MCA, we proposed that the vascular abnormalities caused by SV in the left MCA destroyed the lenticulostriate arteries, leading to ischaemic lesions in the left basal ganglia. HR-VWI is a valuable imaging tool that provides information on affected arteries, and in our case, it provided nuanced change information about arterial wall integrity and pathology.

Because SV often lacks typical symptoms of syphilis, like fever and rash, it is difficult to diagnose with clinical manifestations alone. In this regard, the HR-VWI characteristics of SV can help physicians distinguish the rare aetiology of intracranial vascular stenosis. Characteristics of SV in HR-VWI are mainly associated with concentric wall thickening and circular uniform enhancement,⁴ while characterisation of atherosclerosis disease in HR-VWI is mainly associated with eccentric, irregular wall thickening and focal uneven enhancement.⁵ The HR-VWI showed the concentric thickening and contrast enhancement in our case. Of course, the concentric thickening and contrast enhancement in HR-VMI more often occurred in patients with central nervous system (CNS) vasculitis. CNS vasculitis and SV often cause multiple lesions in the intracranial vessels such as the Willis circle, basilar artery, or lenticulostriate artery. When the intracranial vascular presented as only isolated local stenosis or occlusion, the causes were more difficult to determine. Our case presented as isolated intracranial vascular stenosis, so we needed stronger evidence to prove that the SV was the main cause of the MCA stenosis. The HR-VWI changes of the isolated stenosis artery can help to identify SV.² The follow-up findings of our case demonstrated that HR-VWI showed a significant reduction in the degree of wall thickening and stenosis and the enhancement after anti-syphilis treatment with antiplatelet drugs. CNS vasculitis cannot be treated without steroids or immunosuppression treatment. Therefore, it

proved that the anti-syphilis treatment was effective and the cause of isolated middle cerebral artery stenosis in our case was SV. Indeed, the main limitation of our case was that we cannot obtain the pathological result to confirm the clinical diagnosis.

Although concentric wall thickening and circular enhancement of the intracranial vessel wall on HR-VWI are not specific manifestations of SV,⁶ the changes in the HR-VWI images after anti-syphilis treatment can help confirm the diagnosis of SV. From the initial concentric wall thickening and circular contrast enhancement to the reduction of wall thickness and the disappearance of enhancement effect after anti-syphilis treatment, HR-VWI is a useful method to verify suspected cases of SV and could serve as a biomarker for assessing the response to anti-syphilis treatment in the future.

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Data availability. The datasets used during the present study are available from the corresponding author on reasonable request.

Conflicts of interest. The authors declare no conflicts of interest.

Declaration of funding. This study was funded by Deep Blue 123 key project of The First Affiliated Hospital of Naval Military Medical University (2020YSL004).

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