Atypical Radiographic Presentation of Multiple Sclerosis with Bilateral Internal Jugular Vein Occlusions

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Background
Chronic cerebrospinal venous insufficiency (CCSVI) is a hypothesis that attempts to link alarmed cerebrovascular venous outflow with the pathogenesis of MS.1 There have been conflicting studies on this topic, and recent work has identified extracranial and intracranial venous stenosis and occlusions in MS patients and healthy controls with a similar frequency.2

The initial studies utilized Doppler sonography to identify abnormalities; however, MR venography has been suggested as a sensitive, widely used, and operator-independent tool for evaluation of the cerebrovascular tree.3 The presence of CCSVI in MS has not shown a significant correlation with degree of clinical disability.4 It is not known whether venous anomalies might influence the radiographic appearance and lesion distribution in MS.

Objectives
We describe the clinical and radiographic findings of a patient who presented with a clinical syndrome suggestive of MS and bilateral internal jugular vein occlusions.

Case Report
- 51 y.o. man developed monocular visual impairment followed by impairment in the other eye within one month. Recovery was near complete, with residual Uhoff’s phenomenon.
- He developed a sensory myelopathy several weeks later, followed by depression, memory loss, and worsening difficulty.
- MRI of the brain demonstrated multiple T2/FLAIR hyperintensities, many enhancing, predominantly involving the subcortical U-fibers in the posterior left hemisphere (Figure 1). There were lesions in the left pons (Figure 2A), right medulla, right cerebellum, corpus callosum, and periventricular regions.
- MRI of the spine revealed lesions at T12/L1 and an enhancing lesion at T2/L1 (Figure 2B).
- CSF findings: 3 oligoclonal bands; Laboratory screening for systemic autoimmune and connective tissue disease was negative.
- Visual evoked potentials abnormal bilaterally.
- MRI/MRV was obtained to rule out venous disease and demonstrated bilateral internal jugular vein occlusions with prominent collaterals in the neck (Figure 3).
- Symptoms responded to a brief course of corticosteroids.
- Follow-up MRI 3 months later demonstrated resolution of enhancement and no new lesions.

Figure 1: Brain MRI

Figure 2: Brain and Thoracic Spine MRI

Figure 3: MR Venogram

Conclusions
- It is not clear whether the finding of bilateral jugular vein occlusion was related or correlated to the acute brain lesions or symptoms in this patient, and if related, whether this was caused an inflammatory demyelinating disease or other pathology (e.g. vasculopathy or a tumor) favored by CSF or venography.
- The vascular findings might also be incidental. The presence of a well-developed collateral drainage system in the neck argues for a chronic process with compartmental outflow.
- Our patient’s MRI findings were atypical, but consistent with MS. Exclusive involvement of the subcortical U-fibers within a single section in patients with MS has been reported.5
- Further research may help to clarify whether there are characteristic and possibly atypical radiographic appearances of demyelination in the subset of patients with various anomalies.

References
2. Weisbrod R et al. Association of abnormal cerebrospinal venous outflow with multisystem involvement a magnetic resonance venography and the qualification study. J Neurol Neurosurg Psychiatry 2011;82:1252-6
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Figure 3: Narrowing and complete or near-complete occlusion of the internal jugular vein (arrows). Enlarged venous collaterals in the neck (arrows).