Introduction— Neurological manifestations of Behçet’s disease (BD) account for 5.3 to 59% of cases, and can be significantly disabling. Their management is not well codified.

Methods— A retrospective analysis of 104 patients with parenchymal neurological involvement of BD (neuro-BD). All patients fulfilled the International Criteria for BD. The disability status was assessed using the Rankin score.

Results— The mean (SD) age at onset was 38.0 ± 11.8 years, and the male to female ratio was 1.3. BD manifestations included oral (104/104, 100%), genital (75/104, 72.1%) ulcerations, ocular (66/104, 63.5%), articulat (50/104, 48.1%) and vascular involvement (31/104, 29.8%). An acute onset accounted for 73/104 (70.2%) of neuro-BD patients. Main symptoms at diagnosis of neuro-BD included headache (n = 65, 62.5%), pyramidal signs (n = 52, 50%), ataxia (n = 27, 26%), and tremor (n = 27, 26%). Meningitis was present in 67/104 (64.4%). The median (IQR) Rankin score at onset was 2 (2–3). The reparation of inflammatory lesions on MRI included the brainstem (60/91 (65.9%), with extension to the subcortical regions in 21/91 (23.1%)), and isolated capsulo-thalamic (12/91 (13.2%)). Glucocorticosteroids were given in all 104 neuro-BD patients, alone in 17 (16.3%) cases and associated with an immunosuppressant in 87 (83.7%) (cyclophosphamide (n = 45, 43.3%), azathioprine (n = 39, 37.5%), ciclosporin (n = 2, 1.9%) and chloraminophenox (n = 1, 1.0%)). Overall, improvement (>50% of the initial Rankin score) was achieved in 42.3% of patients, partial improvement (0–50%) 26.0%, no improvement in 19.2% and worsening in 9.6%. Median time to clinical efficacy was 1 (1–3) month. Mild to severe sequelae were observed in 52 (50%) patients.

Conclusion— Neurological involvement is a severe manifestation of Behçet’s disease leading to sequelae in half cases. Prompt diagnosis and treatment with glucocorticosteroids associated with an immunosuppressant are mandatory.

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CNS vasculitis

27 Vessel wall characteristics using high-resolution magnetic resonance imaging in reversible cerebral vasoconstriction syndrome and central nervous system vasculitis

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Introduction— Reversible cerebral vasoconstriction syndrome (RCVS) and central nervous system (CNS) vasculitis often have similar initial clinical presentation, laboratory findings and imaging features creating a diagnostic dilemma. High-resolution-3-Tesla Magnetic Resonance Imaging with contrast (HR-MRI) is a non-invasive method to look at intracranial vessel wall characteristics.

Methods— A retrospective analysis of all patients with a diagnosis of RCVS or CNS vasculitis that underwent HR-MRI at our institution was performed. Inclusion criteria for RCVS included acute thunderclap headache with no aerynysmal subarachnoid hemorrhage, normal cerebrospinal fluid and reversible multifocal intracranial vessel stenosis [1]. The Calabrese criteria [2] were used for the primary CNS vasculitis group. Images were reviewed by two radiologists. Demographics, clinical presentation, laboratory testing, imaging studies and outcomes were collected.

Results— Twenty-six patients met inclusion criteria with 13 patients in each group (1 patient had secondary CNS vasculitis due to Varicella Zoster). Median age was 52 and 42 in the RCVS group and the vasculitis groups respectively. Females represented the majority in the RCVS groups 85% (11/13), while only 15% (2/13) were females in the vasculitis group. In the RCVS group, 10/13 had wall thickening, of which four had minimal wall enhancement while remaining six had only vessel wall thickening in the areas of vessel stenosis. Two patients had no vessel wall abnormality and 1 patient had vessel lumen narrowing. In the vasculitis group, 12/13 had vessel wall enhancement as well as wall thickening. Forty-six percent (6/13) of vasculitis patients and 69% (9/13) of RCVS patients had follow up HR-MRI.

Discussion— HR-MRI may be a useful tool in differentiating RCVS from CNS vasculitis, in the acute presentation.

Conclusion— Further studies with larger number of cases are needed to confirm the utility of HR-MRI in the diagnosis of cerebral arteriopathies.

References

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