Neuro-behcet disease: diagnosis & clinical issues and management

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Behçet's disease (BD), is an idiopathic chronic relapsing multisystem vascular-inflammatory disease of unknown origin with oro-genital ulceration and uveitis. The disease affects many organs and systems, including the nervous system.
Clinical and imaging evidence suggests that primary neurological involvement in BD may be subclassified into two major forms: the first one, which is seen in the majority, is characterized as a vascular-inflammatory central nervous system disease with focal/multifocal parenchymal involvement, mostly presenting with a subacute brainstem syndrome and hemiparesis; the other, which has few symptoms and a better outome, is caused by isolated cerebral venous sinus thrombosis and intracranial hypertension, occurring in 10- 20%. These two types rarely occur in the same individual, and their pathogenesis is likely to be different. Isolated behavioral syndromes and peripheral nervous system involvement are rare, whereas a vascular-type headache is relatively common and independent from neurological involvement. Neurologic complications secondary to systemic involvement of BD and related to BD treatments are considered as secondary neurological involvement. The core histopathological phenomenon seems to be a vasculitic involvement in some cases, and low grade chronic non-specific inflammation in others. As the neurological involvement in this syndrome is so heterogeneous, it is difficult to predict its course and prognosis, and response to treatment. Currently, treatment options of NBS are limited to attack therapies with high dose intravenous methylprednisolone followed with a prolonged oral taper and mainly the use of azathioprine, cyclophosphamide, interferon alpha and anti-TNF agents for long term preventive treatment despite no evidence for their efficacy.