A case of central nervous system vasculitis related to an episode of Guillain-Barrè syndrome

ABSTRACT

The authors report their knowledge about an uncommon case of isolated vasculitis, restricted to the left sylvian artery during an auto-immune Guillain-Barrè syndrome (GBS), sustained by cytomegalovirus (CMV). An acute cardiopulmonary failure requiring a ventilator and vasopressor support manifested, notwithstanding plasma exchanging and immune-modulating therapy. An IgM-enriched formula administration coincided with a rapid amelioration of GBS and vasculitis to a complete recovery the next month after her discharge to a rehabilitation centre.

INTRODUCTION

Introduction Isolated vasculitis of the central nervous system (CNS) is an uncommon clinicopathological entity, which is characterized by vasculitis and is restricted to the vessels of the CNS, without any other apparent systemic lesion. Arterioles are the most common target of degenerative processes, with mononuclear cell infiltrates as a distinctive sign, but vessels of any size may be involved. Viruses, such as cytomegalovirus and varicella-zoster virus, bacteria with strong neuronal tropism, such as Treponema pallidum (the agent that is responsible for syphilis), and Hodgkin's disease have been identified as aetiological agents. Herein we report a case of sylvian artery involvement in a young patient affected by an episode of GBS, after coinfection with cytomegalovirus and herpes simplex virus.

CONCLUSION

The mechanism of neurological dysfunction in the vascular system is tissue ischaemia, which produces varied clinical effects, with transient or prolonged symptoms. Scarred vessels, as well as those that are acutely inflamed, are believed to be the main cause of ischaemia. Patients with primary isolated vasculitis of the CNS normally present with very poor signs or neurological defects, but devastating neurological abnormalities may occur depending on the extent of vessel involvement. Mononeuritis multiplex, polyneuropathy and stroke occur with more frequency, but encephalopathies, cranial neuropathies and brachial plexopathies are also seen. It is of primary importance to distinguish between a primary or secondary form of isolated vasculitis of the CNS and a different pathology, and radiology is the best technique to make this distinction. In the past, the only means whereby a diagnosis of isolated vasculitis of the CNS could be confirmed was angiography plus cerebral parenchyma biopsy. Recently, however, some investigators have proposed magnetic resonance "as the most sensitive ancillary procedure to detect CNS damage, being many of the angio-graphic features found in vasculitis non-specific". To our knowledge, this is the first reported case of isolated vasculitis of the CNS secondary to GBS. Some important points must be emphasized in the present report. First, autoimmune antibodies were the cause of both GBS and isolated vasculitis of the CNS. Second, isolated angiitis of the sylvian left artery and its branch was secondary to GBS, and as soon as the patient recovered from the syndrome the neurological manifestations of the disease started to diminish. Third, the persistence of cytomegalovirus and antibodies directed against cytomegalovirus-DNA in the spinal fluid, despite the immune-modulating therapy, can be considered to be the trigger for both pathologies. Finally, resolution of infection with a new

mixture of Igs with high titre of IgM coincided with the beginning of recovery.