



POSTER

Primary angiitis of central nervous system: diagnostic and therapeutic challenges

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Abstract

Primary angiitis of central nervous system (PACNS) is an uncommon vasculitis restricted to spinal cord and brain. Current diagnostic criteria for PACNS require newly acquired neurological deficit; specific angio-graphic and/or histopathological features of angiitis within the CNS; and no evidence of an underlying systemic disorder. In clinical practice, PACNS is frequently a diagnostic and therapeutic challenge. A 56-year-old man with a past medical history of hypertension and left lobar intracerebral haemorrhage, was admitted with an acute cognitive impairment. A brain MRI revealed multiple acute and chronic brain ischaemic lesions on different vascular territories. Digital subtraction angiography showed bilateral PCA and left ACA occlusions as well as multifocal segmental stenosis of both MCAs. CSF analysis revealed mild pleocytosis and elevated protein. A second

brain MRI showed new subclinical posterior circulation ischaemic lesions with DWI restriction. Additional cardiovascular and systemic autoimmune workup were unremarkable. After brain biopsy we started five days of methylprednisolone followed by oral prednisolone. Although brain biopsy was negative for vasculitis, we maintained corticosteroids and added azathioprine. After three months of follow-up the patient remains clinically stable. A typical clinical picture and specific angiographic features of PACNS could be associated with a negative brain biopsy. Several questions remain unanswered and should be discussed. Can we skip brain biopsy before starting treatment? Should we maintain treatment in negative biopsy PACNS? Which is the best treatment for PACNS?

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