

Primary angiitis of the central nervous system (PACNS)

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In this case report we present a patient with cognitive impairment as a result of primary angiitis of the central nervous system (PACNS). Our patient is male, age 54, with 15 years of education. Patient presented with difficulty in pronouncing, nasal speech and weakness during period of one year. Neurocognitive status showed impairment of episodic memory and concentration, attention verbal, and visual memory deficiency (MMSE 25). Brain CT scan showed no acute hemorrhage or ischemia, but chronic vascular changes in deep white matter of cerebral hemispheres, basal ganglia, thalamus, and pons with lacunar lesion next to the head of left nucleus caudatus. Brain MRI showed intracerebral hemorrhage in right temporal lobe, multiple gliotic/malacian zone paraventricular, predominant in basal ganglia and hemispheres of the cerebellum. Differential diagnose of multiple intracerebral hemorrhage includes vascular malformations, cerebral amyloid angiopathy, ischemic stroke, cerebral venous sinus, tumor, cerebral vasculitis etc.

Medical diagnostic included laboratory tests, prostigmin test, EMNG, AchR and MuSK antibodies test, EEG, cerebrospinal fluid analysis. Through his medical evaluation, which has been performed to exclude any infections or malignant process, we finally conducted cerebral digital subtraction angiography (DSA) to diagnose PACNS. Cerebral DSA showed multiple, non significant stenosis on specific vascular segments, implying high suspected vasculitis. After immunologist being consulted, patient was prescribed with methylprednisolone during 3 next days and cyclophosphamide next 6 months. Control DSA is planned in the future. PACNS is one of the most formidable diagnostic and therapeutic challenges to neurologists. Given the low specificity of cerebral angiogram and not uniform clinical presentation, PACNS is best approached by an organized team with expertise in neurovascular disease, immunology or rheumatology, neuroradiology, and neuropathology.

Keywords: cognitive, vasculitis, DSA, hemorrhage

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