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Primary angiitis of the central nervous system a riddle, wrapped in a mystery, inside an enigma

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54-year-old gentleman with a previous medical history consistent of recurrent headaches, hypertension, dilated cardiomyopathy, hypercholesterolemia and recurrent renal calculi. Over a one-year period, he gradually developed worsening headaches associated with intermittent blurred vision, generalized aches and pains, mild cognitive impairment, and several episodes of focal upper and lower limb weakness. These symptoms led to several evaluations in the A&E and EADU, one evaluation in the TIA clinic, two admissions to the stroke ward and several outpatient reviews in our stroke, neurology and rheumatology clinic. He received a diagnosis of temporal arteritis in the TIA clinic and started to take steroids. He was then admitted to our stroke ward following a diagnosis of multi-territory brain infarctions (pontine stroke, corpus callosum and right centrum semiovale). Following discharge, he remained symptomatic and this led to several outpatients MRIs which revealed more ischemic events disseminated in time and place; however, due to an rather atypical 'resolution' of previous ischemic events, this led to the suspicion of a possible case of multiple sclerosis, leading to an lumbar puncture which ultimately was slightly abnormal but was not positive for oligoconal bands. In the meantime, all serologic investigations were negative for infectious, autoimmune and neoplastic causes. Finally, a CT-angiogram followed by a Digital Subtraction Angiogram (DSA) revealed – in comparison with a previous CTA – a considerable narrowing in both MCAs, leading to the diagnosis of primary angiitis of the central nervous system (PACNS). After IV methylprednisolone, the patient stabilized for a short period of time, only to have a large L-MCA stroke which required a decompressive hemicraniectomy; he remains globally aphasic with mild right upper limb weakness.

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