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Microaneurysms in Primary Angiitis of the Central Nervous System Revealed by MRI

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Background

Primary angiitis of the central nervous system (PACNS) is a rare challenging diagnosis evoked in front of heterogeneous clinical and radiological signs, often leading to invasive diagnostic procedures such as conventional cerebral angiography or brain biopsy [1, 2]. Although still poorly understood, a few clinical, radiological and histological patterns emerged from the analysis of case reports and series of patients with PACNS over the past few years [1, 2]. Recent progress in MRI could be helpful by improving noninvasive early diagnosis accuracy, demarcation of homogeneous subgroups of patients, and physiopathology understanding in PACNS. We report a single case of PACNS with intracranial multiple microaneurysms firstly suspected and followed under treatment on brain 1.5-T-MRI 3-D-susceptibility weighted angiography (SWAN), a new method for susceptibility-weighted imaging with a three-dimensional multi-echo gradient echo MRI providing both higher sensitivity and better spatial resolution than conventional T2-gradient-echo-MRI (T2*).

Summary of Case

A healthy 37 year-old-female suffered from a 5-min lasting right hemiface paresthesias and palsy after an unusual short run, diagnosed as a transient ischemic attack. Physical examination was normal. Brain 1.5 T-MRI revealed disseminated infra- and supratentorial white-matter lesions (fig. 1a) hyperintense on fluid-attenuated inversion recovery and diffusion-weighted imaging with slightly increased apparent diffusion coefficient ratio of which most were enhanced on post-gadolinium T1-WI; T2* was unremarkable, while SWAN revealed six additional supra-tentorial hypointense lesions within multiple cortical sulci (fig. 1b). Hypointense lesions on SWAN were hypothesized as being whether sulcal or cortical hemosiderin deposits (microbleeds), or microvascular very small arterial or venous dilatations, after localized calcifications were excluded on CT-scan. A cerebral angiitis was thus hypothesized and confirmed on conventional angiography (fig. 1c) showing 6 arterial microaneurysms, of which distribution within distal arterial branches and size (maximum diameter ranged from 2 to 7 mm) after attentive comparison by radiologist seemed to match well with hypointense lesions seen on SWAN, and two additional smaller microaneurysms, associated with segmental narrowing and fusiform dilatation within distal branches of vertebral, anterior, and middle cerebral arteries, typical of arteritis. Cerebrospinal liquid analysis showed a lymphocytic pleiocytosis with no oligoclonal bands. After a negative extensive work-up, the diagnosis of primary angiitis of the central nervous system (PACNS) was finally retained. Intravenous methylprednisolone for three days and cyclophosphamide pulse for 3 months followed by oral azathioprine and prednisone enabled early and sustained remission (36 months of follow-up) and dramatic improvement on MRI with a net decrease of hypointense lesions size on SWAN over two years before to stabilize (fig. 1d, e).

This case illustrates a form of aneurysmal PACNS [1–4], without any subarachnoid or parenchymal hemorrhage at presentation or during follow-up. Aneurysms or microaneurysms have been occasionally reported in association with other typical angiographic findings in PACNS but was not systematically described in the largest cohort ever reported so far, and were thus considered being very rare until recently, although considered being specific of arteritis [1–5]. Among this cohort including 131 patients with PACNS [2], Salvarany and colleagues, however, mentioned 3 patients out of 16 with cerebral hemorrhage wearing intracranial aneurysms, localized within the intracranial carotid artery and associated with subarachnoid hemorrhage in two cases [6]. PACNS with intracranial hemorrhage could be related to necrotizing histologic pattern [6], but analysis of histology among patients with PACNS-associated aneurysms is lacking so far. More recently, among the French cohort 'COVAC,' which studied 52 patients with PACNS, including the patient we have described here, 5 patients (13%) were found with intracranial microaneurysms on conventional angiography, which are thus probably not so rare than previously thought [7]. If patients with PACNS-associated microaneurysms represent a specific subgroup with histologic correlation and prognosis implication remain unknown and deserves to be further studied. SWAN highly contributed both to support diagnosis of cerebral angiitis and to follow-up under therapy in this case, since revealing abnormalities into small cerebral vessels, presumably microaneurysms, or superficial small microbleeds, suggestive of cerebral arteritis in this context. Cortical or subcortical petechial hemorrhages have been previously described with T2* in PACNS [8], while noninvasive visualization of microaneurysms with MRI had never been reported so far. Given the higher sensitivity of SWAN in hemosiderin deposits detection compared to T2*, we can also hypothesize that small hypointense lesions on SWAN in our patient may correspond either to such small petechial hemorrhages resulting from inflammatory microvessels leakage or to small venous dilatations related to vasculitis.

Conclusions

We report an original and illustrative case of PACNS-associated microaneurysms firstly suspected on SWAN-MRI. 3-D-susceptibility weighted angiography may offer a new opportunity for im-

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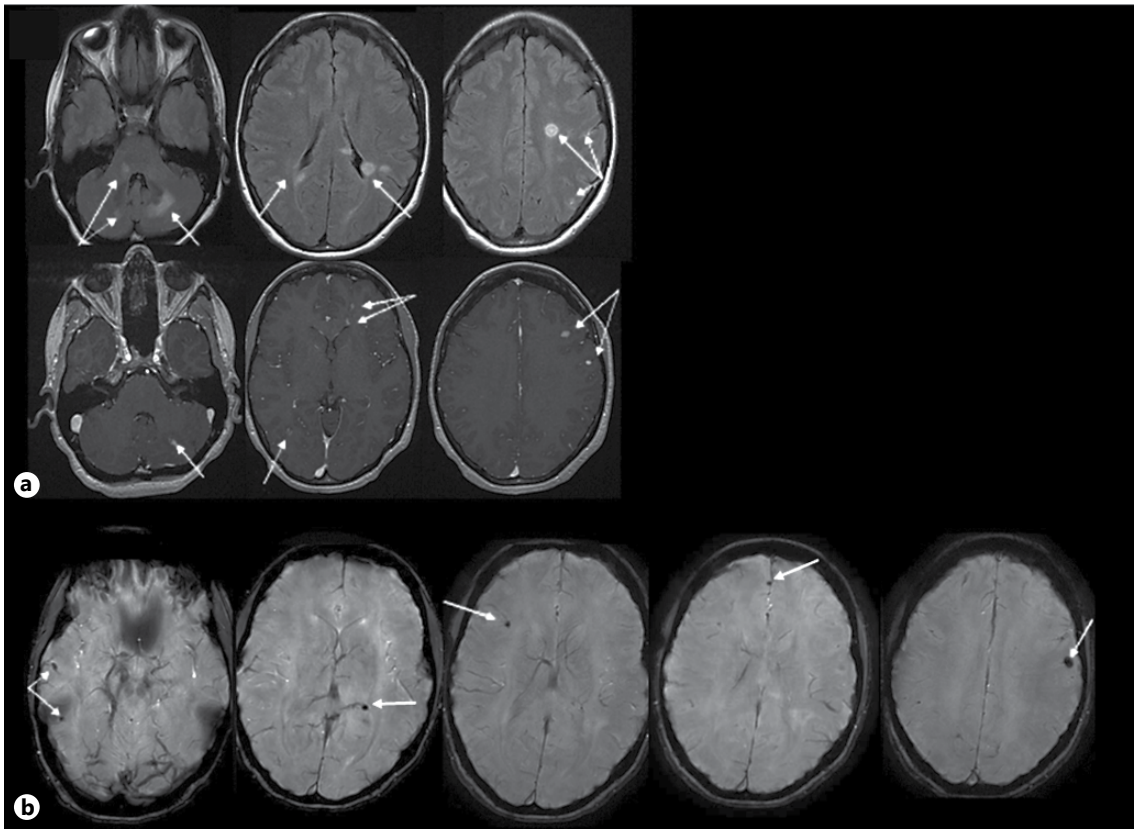

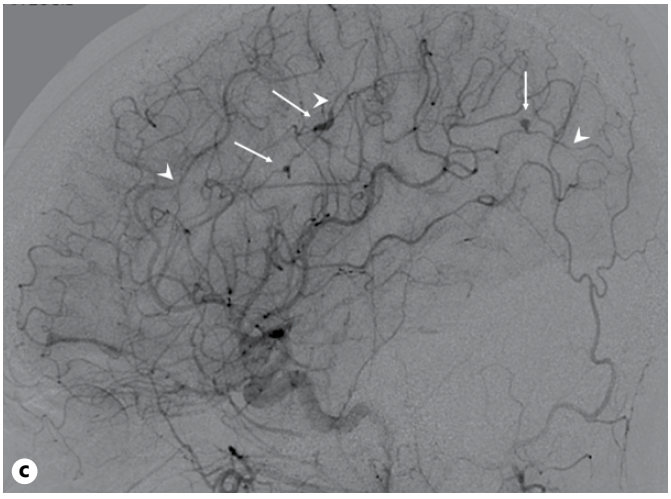

Fig. 1. a Brain 1.5 T-MRI seven days after symptoms onset: Diffuse multiple white-matter lesions (arrows) hyperintense on axial fluid-attenuated inversion recovery (top) or enhanced on post-gadolinium T1-WI (bottom). **b** Brain 1.5 T-MRI seven days after symptoms onset: six sulcal hypointensities on axial 3D-susceptibility weighted angiography (SWAN) within bilateral frontal and temporal convexity (arrows). **c** conventional angiography showing fusiform microaneurysms (arrows) and segmental narrowings (arrowheads) within distal left middle cerebral artery branches.

proving noninvasive diagnosis and follow-up in cerebral arteritis (including PACNS), and should thus be considered in multimodal-MRI protocol when cerebral angiitis is evoked, although confirmation on conventional angiography still remains necessary.

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I, Nicolas Gaillard, in the name of all coauthors, certify that all coauthors have seen and agreed with the contents of the manuscript and provided written permission that the manuscript conforms to ICMJE requirements and presents honest work. I certify that this submission is not under review and its contents have not been reported in publications so far. The patient described in this submission was included in the prospective French COVAC cohort, results of which have been published recently.

Sincerely, Dr. NG.

Dr. Nicolas Gaillard: took part in design and conceptualization, analysis of the data and drafting the manuscript. Dr. NG reports no disclosure.

Dr. Jean Louis Bertrand: took part in interpretation of the data. Dr. JLB reports no disclosure.

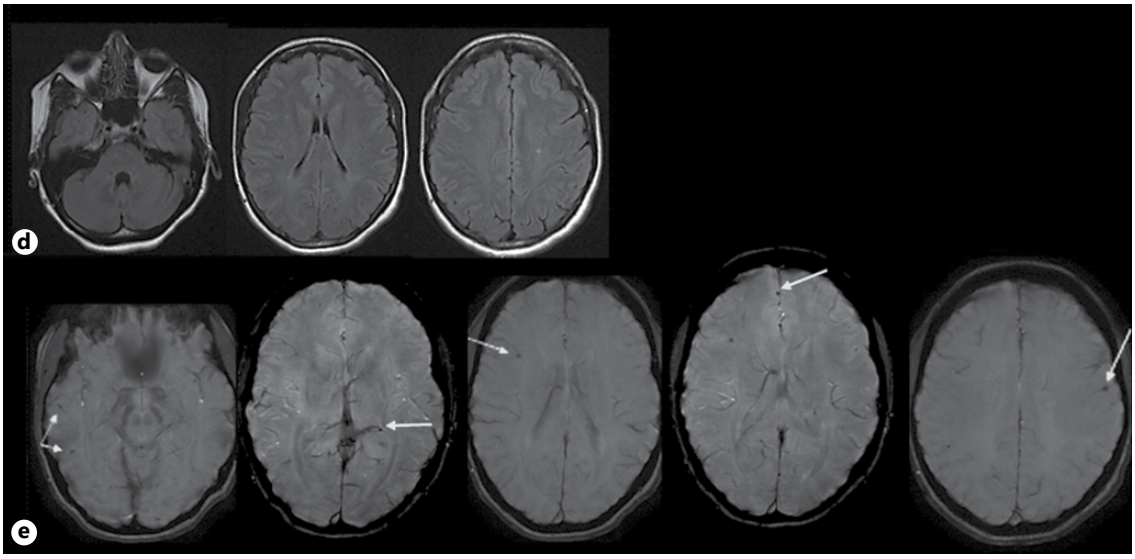

Fig. 1. d Follow-up MRI after 2-years showing almost disappearance of lesions on FLAIR. **e** Follow-up MRI after 2-years showing net size decrease of hypointense lesions on SWAN.

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None.

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