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Histopathologic data on basal cerebral artery alterations in large vessel vasculitis of the brain are scarce. However, several recent reports suggest that these alterations may be noninvasively visualized by MRI. Specifically, high-resolution, fat and blood signal suppressed (black-blood) T1-weighted imaging has demonstrated contrast enhancement in the wall of affected arteries.1,2 In the presented case, we were able to correlate these MRI findings with conventional angiography and histopathology.

Case report. A 59-year-old otherwise healthy woman without apparent vascular risk factors initially presented with moderate right-sided hemiparesis. Diffusion-weighted MRI revealed small acute left frontal brain infarction. Digital subtraction angiography (DSA) demonstrated high-grade stenosis of the terminal segment of the left internal carotid artery (ICA) and the proximal portions of the left middle (MCA) and anterior cerebral artery (ACA) (figure, A). High-resolution black-blood T1-weighted MRI demonstrated marked contrast enhancement in the terminal segment of the left ICA and the left posterior communicating artery (PCoA) (figure, B and C). Repeated CSF examinations showed mild pleocytosis with 7 to 19 cells/mm³ (mainly lymphocytes and monocytes) and a normal protein content and glucose CSF/serum ratio. Erythrocyte sedimentation rate (ESR), leukocyte count, and C-reactive protein were normal in repeated testings. Serology was negative for HIV, viral hepatitis, chlamydia, Treponema pallidum, and Mycoplasma pneumoniae infection. CSF testing was negative for varicella zoster virus, Epstein-Barr virus, cytomegalovirus, Borrelia burgdorferi, and mycotic disease. Extensive laboratory testing (ANA differentiation, ANCA, C3, C4, cryoglobulins, ds-DNA), dermatologic and ophthalmologic examinations, as well as CT of the thorax, abdomen, and pelvis revealed no evidence of systemic vasculitis or malignancy.

A diagnosis of primary angiitis of the CNS (PACNS) affecting the basal cerebral arteries was made and treatment with daily oral doses of 100 mg aspirin, 20 mg simvastatin, 80 mg prednisolone (later tapered to 20 mg), and 100 mg cyclophosphamide (later reduced to 50 mg) was initiated. Eight months later, the patient developed progressive left hemispheric infarction despite therapy. Immunosuppression was immediately escalated to IV methylprednisolone (1 g) and IV cyclophosphamide (1 g/m² body surface area). However, within 2 days, infarction had extended to the complete left anterior and middle cerebral artery territory. Another 4 days later, isolated brain death due to malignant infarct swelling was ascertained.

Postmortem examination of the brain revealed a massively swollen brain with infarction of the left MCA and ACA territory and a severe midline shift to the right. Histology of the basal cerebral arteries revealed severe vasculitic alterations in the terminal segment of the left ICA, the proximal portions of the left MCA, ACA, and posterior cerebral artery (PCA), the left PCoA, as well as the proximal portion of the left superior cerebellar artery (SCA). Other extracranial or intracranial arteries showed no involvement. In the affected arteries, extensive thickening of the vessel walls and narrowing of the lumina (figure, D), fibrinoid necrosis of the media, blurring boundaries between media and adventitia, as well as lymphocytic infiltration of the intima (figure, E) could be seen in hematoxylin-eosin stains. Furthermore, multinucleated giant cells of foreign body type were detectable in high-power magnifications (figure, F).

Discussion. The diagnosis of PACNS is difficult and depends on the presence of angiographic or histopathologic evidence of angiitis in the absence of systemic vasculitis.3,4 Moreover, PACNS has to be distinguished from reversible cerebral vasocostriction syndrome, a more benign syndrome with a variety of different triggers and a self-limiting course.5 Since PACNS may affect different calibers of arteries in the brain, a classification in large, medium, and small vessel vasculitis has been sug-
gested. According to this classification, our case qualifies as large vessel PACNS restricted to the basal cerebral arteries. Histopathology demonstrated lymphocytic infiltration and multinucleated giant cells of foreign body type. The absence of extracranial artery involvement and a normal ESR argue against a diagnosis of temporal arteritis or Takayasu arteritis. The constellation in the presented case may suggest another distinct manifestation of giant cell arteries with predilection of the basal cerebral arteries. Further studies are needed to confirm that the observed pattern may be typical for some cases of large vessel PACNS and that black-blood T1-weighted MRI may be valuable in the diagnosis of this and other neurovascular diseases in which vessel wall inflammation may play a role.

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