Abstract: The purpose of the study was to examine the value of the non-invasive magnet resonance angiography (MRA) in the follow-up of cerebral vasculitis (CV) and vasculitis-like angiopathy. We performed follow-up MRA (TOF 3D), MRI and transcranial doppler ultrasound (TCD) in the patients with isolated angiitis of the CNS (2/6), Crohn-disease-associated CV (1/6), and reversible arterial vasoconstriction (RAV) of the CNS (1 migraine, 1 eclampsia and 1 toxic encephalopathy) (3/6). In all patients with RAV MRA showed a complete remission of the vascular alterations after treatment. In the patients with isolated angiitis of the CNS and Crohn-disease-associated CV, partly regressive and partly progressive changes were demonstrated. The MR-angiographically detectable vascular alterations corresponded to the clinical course of the disease, as well as to TCD in all our patients. Success of therapeutic procedures, the need and the intensity of further drug administration could be estimated. The MRA appears to be a valuable non-invasive method in the follow-up of patients with CV and RAV.

Key words: magnetic resonance angiography; cerebral vasculitis; reversible arterial vasoconstriction

INTRODUCTION

Cerebral vasculitis and the heterogeneous group of reversible arterial vasoconstriction (RAV), also called benign angiopathy or vasospasm-caused angiopathy of the central nervous system (CNS), are well-known as diseases that produce serious neurological symptoms. Whereas the RAV is usually associated with neurological or non-neurological diseases with clear clinical features as migraine or eclampsia, cerebral vasculitis imposes a great diagnostic challenge because of unspecific clinical signs and a lack of efficient non-invasive diagnostic modalities [7, 8].

Although catheter angiography is acknowledged as method with good sensitivity for the demonstration of vascular alterations in cerebral vasculitis and RAV [8, 9, 18], the invasive character of this examination, false negative results in about 25 %, and a low specificity reduce the acceptance as gold standard [8, 11]. Furthermore, particularly the ischemic complications of catheter angiography reduce the acceptance of this method for follow-up investigations.

First positive experiences with high resolution magnet resonance angiography (MRA) for detection of cerebral vasculitis have been published in the last few years [5, 21, 28, 29]. In this study we examined the value of MRA in the follow-up of several patients suffering from cerebral vasculitis or RAV.

METHODS

Six patients (age range 18-60 years; 6 female) were diagnosed by clinical, laboratory and radiological methods to suffer from isolated angiitis of the CNS (3/6), Crohn-disease-associated CV (1/6), and RAV of the CNS (1 migraine, 1 eclampsia and 1 toxic encephalopathy) (3/6). Other inflammatory (particularly systemic vasculitis), neurodegenerative, metastatic or embolic diseases were excluded in these patients.

MRI (T2w-TIRM, T2w-FLAIR, T1w-SE, T1w-SE with Gadolinium), and MRA (TOF 3D FISP) with an 1.5 T MRI-scanner (Siemens Magnetom Vision and Sonata, Erlangen, Germany) and transcranial doppler ultrasound (TCD) were performed initially in all 6 patients, and digital subtraction catheter angiography (DSA) in 4 patients within 48 hours after diagnostic MRA (Tables 1 and 2). DSA was not performed in 1 patient with typical migraine without aura and in 1 patient with classical eclampsia (Patients 1 and 3, Table 1). Additional diffusion-weighted MRI was performed in 4 patients. Follow-up MRA and MRI were performed within a mean interval of 15 days (+ 6.8). Two patients received a second, and one patient a third follow-up investigation. TOF 3D FISP for MRA was performed by use of a circular polarised head coil with the following parameters: TR 35 ms; TE 7.2 ms; Flip Angle 25°; band width 81 Hz/pixel; voxel size 0.75 mm x 0.39 mm x 0.75 mm; duration 6:44 min. Axial and frontal rotation of MIP images followed data reconstruction. In addition we performed MRI by use of T2w TSE (spectral fat saturated), T2w FLAIR, T1w SE before and after injection of gadodiamid (Nycomed, Ismaning, Germany).

Focal or diffuse vessel narrowing, ectasia, string-of-bead-sign, or complete loss of depiction of first and second order cerebral vessels were defined as...
MRA criteria for pathological changes of these vessels. Because of the well-known artefacts in MRA due to turbulent flow and saturation effects, vessels beyond 2nd order were not evaluated. Furthermore, inhomogeneous intravascular signal in conjunction with bad signal-to-noise ratio was considered indeterminate and not taken to represent vessel pathology. Homogeneous, hour-glass like and smooth narrowing or loss of intravascular signal were taken as indicators for real vessel lesions. TCD was interpreted considering established sonographical criteria of pathologically accelerated blood flow.

The results of both initial and follow-up radiological investigations were compared with the clinical course of the disease in each patient.

**RESULTS**

The clinical symptoms, MR-tomographical, MR-angiographical, X-ray angiographical (DSA), and TCD-changes are presented with Tables 1 and 2. Clinical and radiological investigations performed at the same day are presented with numbers as the first, second or third investigation in the Tables.

Complete MR-angiographical remission of vascular alterations after treatment could be found in all 3 patients with RAV. Narrowing of multiple branches of cerebral arteries in the patient 1 with migraine (Fig. 1), as well as stenotic lesions of the anterior and middle cerebral arteries (ACA, MCA) in the patient 2 with toxic encephalopathy (both without parenchymal changes in MRI) could not be demonstrated in the follow up MRA (Fig. 2).

The initial narrowing of the posterior and middle cerebral arteries (PCA, MCA) in our patient 3 with eclampsia were also not demonstrated in the follow up investigation (Fig. 3, Table 1).

Decrease of the length, amount and quantity of stenosis under immunsupressive medication could be shown in one patient with isolated angiitis of the CNS (patient 4) (Fig. 4).

In another patient with isolated angiitis of the CNS (patient 5) with late start of therapy and fluctuating focal neurological symptoms progressive parenchymal lesions in MRI compatible with MR-angiographically only slightly regressive vascular changes even in third follow-up 146 days after the first investigation were found (Table 2).

In our patient 6 with Crohn-disease-associated CV follow-up MRA demonstrated a progression of vessel irregularities and narrowing of cerebral arteries, concurring to the progression of parenchymal lesions (Fig. 5). A 2nd follow-up MR-investigation after improvement of the clinical symptoms in this patient was not performed, the regression of initially accelerated blood flow velocities could be, however, demonstrated in TCD (Table 2).

**DISCUSSION**

The heterogeneous disease group of cerebral vasculitis and reversible arterial vasoconstrictions needs a prompt diagnosis and a sufficient treatment to avoid severe complications. Clinical improvement of neuro-
<table>
<thead>
<tr>
<th>Patient/Age/Sex</th>
<th>Diagnosis</th>
<th>Main clinical presentations</th>
<th>MRI (T2)</th>
<th>MRA</th>
<th>DSA</th>
<th>TCD</th>
</tr>
</thead>
<tbody>
<tr>
<td>4. PS/29/f</td>
<td>isolated cerebral angiitis</td>
<td>1. headache, left hemiparesis, hemihypesthesia, and visual field defect</td>
<td>1. multiple confluent edemas, infarctions in territories of right PCA and MCA</td>
<td>1. multiple narrowing</td>
<td>distinct right PCA stenosis, irregularities left MCA, loss left distal PCA depiction</td>
<td>1. multiloculary increased blood flow right ACI, MCA, PCA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. progressive left hemiparesis</td>
<td>2. regressive hyperintensities in territories of right PCA, progressive in territory of right MCA regressive signal in territories of right PCA and MCA</td>
<td>2. progressive narrowing of right PCA and proximal branches of MCA</td>
<td></td>
<td>2. multiloculary accelerated blood flow right ACI, MCA, PCA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. regressive left hemiparesis</td>
<td>3. regressive right PCA narrowing, irregularities right MCA</td>
<td></td>
<td></td>
<td>3. regressive blood velocities</td>
</tr>
<tr>
<td>5. LB/46/f</td>
<td>isolated cerebral angiitis</td>
<td>1. headache, left arm paresis</td>
<td>1. multiple lesions of both ACA and MCA</td>
<td>1. loss right ACA depiction, loss left MCA</td>
<td></td>
<td>1. accelerated blood flow velocities left and right ACA and MCA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. headache, fluctuating left hemiparesis</td>
<td>2. slightly progressive infarctions</td>
<td>2. identical changes</td>
<td></td>
<td>2. similar changes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. fluctuating left hemiparesis</td>
<td>3. slightly progressive infarctions, newly infarctions left MCA</td>
<td>3. slightly regressive right PCA narrowing</td>
<td></td>
<td>3. similar changes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4. headache, fluctuating left hemiparesis</td>
<td>4. slightly progressive infarctions</td>
<td>4. identical changes</td>
<td></td>
<td>4. similar changes</td>
</tr>
<tr>
<td>6. WK/41/f</td>
<td>Crohn-disease-associated cerebral vasculitis</td>
<td>1. confusion, aphasia, headache, right hemiparesis</td>
<td>1. older small infarctions no new lesion</td>
<td>1. multiple narrowing of both ACA and MCA, and right PCA</td>
<td>multiple narrowing of both ACA, MCA, and right PCA</td>
<td>1. accelerated blood flow velocities both ACA, MCA and PCA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. progressive hemiparesis, right hemianopsia</td>
<td>2. progressive multiple infarctions both MCA, left ACA, right PCA</td>
<td>2. progression of multiple narrowing</td>
<td></td>
<td>2. identical changes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. regressive hemiparesis</td>
<td></td>
<td></td>
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<td>3. regressive blood flow velocities</td>
</tr>
</tbody>
</table>
Fig. 1. 1.1. Initially performed MRA of patient 1 shows multiple narrowing of the right MCA; 1.2. The follow-up investigation of patient 1 demonstrates normal MRA without narrowing.

Fig. 2. 2.1. Initially performed DSA of patient 2 with no irregularities of vessels; 2.2. Initially performed MRA of patient 2 demonstrates multiple narrowing of the left MCA and both ACA; 2.3. Follow-up MRA of patient 2 two weeks after first diagnostic MRA demonstrates almost complete normal vessels with residual narrowing of trifurcation of the left MCA.
logical symptoms under therapy suggests, of course, an effectiveness of the medication [6, 20]. However, a control of vascular changes is desirable to evaluate the local remission effect. Reproducibility and comparability are required from an imaging modality suitable for follow-up. MRI and MRA offer these opportunities and enable examination of parenchymal and vascular structures without invasive character.

Concurring to angiographical studies [13, 23], the attack-associated multifocal cerebral segmental vasoconstriction in our previously described [30] patient 1 with common migraine was completely transient, as well as the multifocal vessel narrowing of cerebral arteries in the patient 2 with toxic encephalopathy. Interestingly, the RAV that has been reported with use of cocaine, amphetamines, heroin and LSD [32] was not previously observed in intoxication with self-made alcohol drinks as in our patient 2. The multifocal vessel narrowing in MRA in this patient that correlated with results of the TCD and were well compatible with focal neurological symptoms, could not be, however, shown in the DSA performed 48 hours after MRA and TCD. The transient character of the vasospasm and immediately begin and effect of the
treatment could be the causes of this inconsistency in MRA and DSA. Patient 3 with eclampsia had also a complete MR-angiographical regression of the arterial narrowing in the PCA and MCA temporally corresponding to the resolving of the clinical symptoms as it was previously demonstrated by others angiographically [10, 26, 33, 35], and with TCD [3, 25], but only once with MRA [31]. Whereas in patients 1 and 2 no edema or parenchymal lesions in the initial MRI could be demonstrated, patient 3 showed a clear regression of the initially demonstrated typically located [19] confluent biparietooccipital vasogenic edema (Table 1).

Catheter angiography is widely used for approval of vascular changing in inflammatory vessel alteration [1, 8, 9, 18]. Sensitivity of about 70 % and specificity of about 30 % lead to a remaining diagnostic insecurity [8, 9]. Even a normal catheter angiography does not exclude an vasculitis or angiopathy. Despite cranial biopsy is the method with the highest specificity (about 75 %) in isolated angiitis of the CNS, a restrictive use of this method is recommended because of the invasive character [15]. MRI is very sensitive for detection of parenchymal lesions in cerebral vasculitis, and a negative MRI excludes intracranial vasogenic edema (Table 1).

Fig. 4. 4.1. Initially performed DSA of patient 4 shows distinct left PCA stenosis; 4.2. Initially performed DSA of patient 4 demonstrates loss of depiction of the right PCA; 4.3. and 4.4. Initially performed MRA of patient 4 show narrowing of left ICA and the right MCA and tandem stenosis of the right PCA; 4.5. Follow-up MRA of patient 4 nine month after first diagnostic MRA demonstrates persisting narrowing of the right PCA, right MCA and left ICA; 4.6. Initially performed T2w Flair Sequence of patient 4 shows edema of right thalamus and right internal capsule corresponding to infarctions of right MCA and PCA; 4.7. T2w Flair Sequence of patient 4 nine month after first investigation demonstrates regressive hyperintensities in the right internal capsule and thalamus.

In our patient 6 with cerebral vasculitis in Crohn disease, the initially MR-angiographically demonstrated narrowing and occlusions in the both MCA and ACA, and in the right PCA could be confirmed with the DSA (Table 2). Although other causes of multiple cerebral ischemias in an acute attack of the Crohn disease as hypercoagulability with raised platelets count and elevated fibrinogen level could not be entirely excluded, there was no echocardiographical evidence of an embolic source in our patient. The finding of multifocal cerebral events in the blood supply territories of various cerebral arteries and multiple segmental narrowing of cerebral arteries in our patient are characteristic for vasculitis of the CNS [9, 11]. The dramatic clinical and MR-angiographical progression despite anticoagulation and the rapid improvement of the symptoms under the treatment with prednisolon in our patient can also be estimated as typical for vasculitis. The MR-angiographical changes in this patient are similar to the vascular alteration demonstrated in DSA in a few cases of Crohn-associated vasculitis of the CNS that have been published so far [2, 4, 14, 22].

High resolution MRA is a routinely applicable examination. First published studies with MRA gave good results for detection of vascular alteration in course of vasculitis [21, 29] and vasospasm caused angiopathy [30]. Present limitations are caused by the reduced spatial resolution of this examination compared with catheter angiography. Peripheral vascular branches can not be evaluated with high accuracy [16, 34]. Negative MRA in patients with parenchymal lesions in MRI does not exclude cerebral vasculitis. However, in cases with positive evidence of vascular alterations by MRA this examination is able to substitute catheter angiography in many cases [29]. Inconsistencies of MRA and DSA in patient 2 (Tables 1 and 2) seem to be related to the reversible course of the disease, because the first diagnostic MRA was
performed 48 hours before the DSA. On the other hand, artefacts as a cause of the inconsistencies can be not entirely excluded. As a whole, concurrence of MR-angiographical changes with MR-tomographical and TCD changes and, particularly, with clinical course could be demonstrated in all our patients. Success of therapeutic procedures, need and intensity of further drug administration could be estimated.

Drawing conclusion we must say that still catheter angiography is the gold standard for assessing intracerebral vessels. However MRA offers a non-invasive method to assess the parenchymal and vascular intra-cerebral changes in patients suffering from vasculitis or vasculopathy.

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