Carotid Artery Stenosis and Ischemic Strokes in Patients with Giant Cell Arteritis

A Characteristic Pattern—Literature Review and Case Report

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Abstract

Purpose Ischemic stroke is a relatively rare complication of giant cell arteritis often accompanied by vessel stenosis. Our purpose was to compare the location of internal carotid artery stenosis in GCA patients by performing a literature review suggesting a specific and characteristic pattern.

Methods We performed a PubMed research including all articles and cited articles reporting cases and case series about giant cell arteritis patients with internal carotid artery stenosis and ischemic strokes.

Results In this case series 39 cases were included. We found a clear tendency of giant cell arteritis-related stenosis to be in the intracranial segments (35/39 (89.7%)). Only in 8/39 (20.5%) patients there was further involvement of extracranial segments. Many cases (27/39 [69.2%]) showed a bilateral involvement.

Discussion This literature review reveals a specific pattern of internal carotid artery involvement in patients with giant cell arteritis and ischemic strokes. To our knowledge this pattern has not been reported as a sign strongly pointing toward giant cell arteritis before. We have not found case reports mentioning other common types of vasculitis reporting this involvement pattern.

Conclusion Internal carotid artery stenosis and ischemic stroke is a rare complication in patients with giant cell arteritis. Considering the characteristic features of bilateral distal internal carotid artery stenosis giant cell arteritis should be suspected which potentially leads to an early diagnosis and immunotherapy.

Keywords
► vasculopathy
► stenosis
► stroke
► vessel wall enhancement
► internal carotid artery
► vasculitis
► imaging
► temporal artery biopsy
► giant cell arteritis
► Horton disease

Introduction

Ischemic stroke in giant cell arteritis (GCA) is a well-known complication. The incidence rate ranges between 2.8 and 7%.1–7 Diagnosis can be challenging due to variable presentation and similarities with other inflammatory vessel diseases or causes of vascular stenosis.

In this literature review we want to discuss the characteristic localization of GCA-related bilateral intracranial internal carotid artery (ICA) stenosis comparing radiological and histopathological findings of previous studies. The possible challenges of diagnosing this disease will be demonstrated.
We want to present our own case of a 58-year-old female patient with suspected temporal-artery-biopsy-negative GCA who suffered recurrent bihemispheric strokes and hemodynamic impairment of both hemispheres while the only manifestation site was both intracranial carotid arteries. Despite immunosuppressive treatment the patient could not be prevented from experiencing new strokes.

**Literature Review**

**Methods**

Our aim was to perform a literature review of cases with internal carotid artery involvement in GCA and ischemic stroke. A PubMed research was performed including articles and cited articles about selected cases of patients describing stenosis of both or one ICA in patients with GCA and ischemic strokes at the time of diagnosis while searching for the terms “giant cell arteritis,” “GCA,” and “carotid artery,” “ischemic stroke” or “intracranial involvement.” The patients had to be diagnosed according to plausible clinical features and current guidelines, by a positive temporal artery biopsy (TAB) or histological proof of giant cells in other large vessels combined with clinical features attributable to GCA. Another criterion was that the segment or segments of stenosis were mentioned or could be figured out by analyzing the provided vascular imaging or autopsy.

Although other signs of inflammation can be drawn to attention we have focused on stenosis, occlusion, or arterial wall thickening revealed by MRA, CTA, conventional angiography, or necropsy. Unless the original site of occlusion could be determined by sonography patients with bilateral proximal ICA occlusion or occlusion of the only involved ICA diagnosed by CTA, MRA or DSA were not patients since the original site of occlusion could be more distal. Moreover, cases with occlusions were considered if a necropsy was performed, and the maximum of inflammatory changes could be identified.

**Results**

This literature review revealed a clear tendency of GCA to cause bilateral intracranial stenoses (mainly cavernous and [para]clinoid segment) in the case of ICA involvement. The patients’ features are shown in Table 1, a summary of the results in Table 2. Bilateral and nearly symmetrical distal occurrence seems to be regular (Fig. 1): in 27/39 (69.2%) cases ICA involvement was bilateral. In four cases there was no information whether stenosis was bilateral. In 35/39 (89.7%) cases there was intracranial (all segments but C1 cervical segment) ICA stenosis. Only in 8/39 (20.5%) cases there was an involvement of extracranial segments. The rate of involvement in relation to the individual segments was C1 cervical segment 8/39 patients (20.5%), C2 petrous segment 5/39 patients (12.8%), C3 lacerum and C4 cavernous segment 27/39 patients (69.2%), C5 (para)clinoid segment 8/39 patients (20.5%), C6 ophthalmic (supraclinoid), and C7 communicating (terminal) segment 12/39 patients (30.8%). In contrast to previous findings, we could not confirm that the previously reported female dominance of the GCA large vessel variant also accounts for ICA involvement (male/female = 22: 13; four cases not specified). A younger age compared with small vessel or only temporal artery GCA patients was apparent: average 68.2 years, median 69 years.

**Case Report**

A 58-year-old female patient was admitted to the emergency room complaining of sudden onset palsy of the left arm and leg. No headache was present. She had a history of hypertension. We diagnosed a diabetes mellitus type II and dyslipidemia. CT angiography and MRI/MR angiography revealed bilateral stenosis of both intracranial ICA pronounced on the right side, a perfusion deficit of the right hemisphere and bilateral new infarcts also pronounced on the right side. A diagnostic cerebral angiography, and MRI showed bilateral infarcts and smooth and mostly concentric bilateral distal ICA stenosis, a 70% stenosis of the right internal carotid artery in the C3 to C6 segments, a 60% stenosis of the left ICA in the same segments, and a partial supply of the right middle and anterior cerebral artery territories by crossflow at the anterior communicating artery (Figs. 2, 3 and 4a). Laboratory investigation revealed an elevated blood sedimentation rate (99 mm/h), C-reactive protein (79 mg/L), and ANA titer (1:640). Antibodies found in rheumatic diseases repeatedly tested negative. CSF was normal. Testing for HIV, VZV, and hepatitis B/C was unremarkable. We discussed primary angiitis of the central nervous system (PACNS) as differential diagnosis but since CSF was normal and BSR and CRP elevated, there was no further involvement of vessels apparent in the conventional angiography other than both ICAs; a brain biopsy was unrevealing and complete vessel occlusions tend to be rare in PACNS. GCA seemed more likely. No other cause for the ischemic strokes, such as atrial fibrillation, could be found. Four months later new infarcts on the right side and an occlusion of the right ICA were found (Fig. 4b). Duplex sonography of cranial arteries according to EULAR recommendations and a whole body PET-CT in search of large vessel involvement (e.g., aortitis) were unrevealing; however, the latter was performed during steroid treatment. A TAB on the right side was performed showing no results of inflammation. A biopsy of the stenotic or occluded carotid artery region was not feasible. Facing progressive ischemic strokes and vessel stenoses we decided to start a steroid treatment suspecting GCA under which BSR and CRP decreased. An MRI (T1 black-blood post-gadolinium imaging sequence) revealed left-sided ICA vessel wall enhancement (VWE) of the cavernous and petrosal segments which gave us a cause to suspect focal arterial inflammation. After treatment with methotrexate, prednisolone and later tocilizumab inflammatory parameters were lowered. However, despite immunotherapy new infarcts occurred (Fig. 6). We decided to present this case although the patient could not be diagnosed with GCA by TAB and the diagnosis is not certain but due to clear signs of inflammation in both distal ICAs there are significant similarities with our case reviews’ characteristic pattern of involvement.
<table>
<thead>
<tr>
<th>Ref.</th>
<th>Neurological symptoms</th>
<th>Age [y]</th>
<th>ESR [mm/h]</th>
<th>Angiography/ Autopsy findings</th>
<th>Extracranial involvement</th>
<th>Intracranial involvement</th>
<th>Bilateral involvement</th>
<th>Diagnosis confirmed by</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>Headache, mild dysarthria, and a left beating horizontal nystagmus.</td>
<td>74/m</td>
<td>61</td>
<td>Focal stenosis in the right carotid siphon (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>Sonography, TAB</td>
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<tr>
<td>33</td>
<td>Headache, Horner syndrome, amaurosis fugax, headache</td>
<td>74/m</td>
<td>n.a.</td>
<td>Moderate stenosis of the right cavernous and supraclinoid internal carotid artery (ICA; white arrow) and, to a lesser extent, the left ICA</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Headache, aphasia, right arm paresis</td>
<td>66/m</td>
<td>n.a.</td>
<td>Both cavernous segment high-grade stenosis</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Headache, gait instability, amaurosis fugax, vision loss</td>
<td>79/m</td>
<td>n.a.</td>
<td>High-grade stenosis of both cavernous/paraclinoid ICA</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Headache, wording difficulties, unsteadiness</td>
<td>59/m</td>
<td>n.a.</td>
<td>Symmetric narrowing in both internal supraclinoid segments</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Right vision loss, hemiparesis</td>
<td>76/f</td>
<td>n.a.</td>
<td>Stenosis of right carotid siphon</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Headache, dysarthria, imbalance</td>
<td>74/m</td>
<td>n.a.</td>
<td>Right proximal and distal cavernous segment stenosis</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>TAB</td>
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<tr>
<td>22</td>
<td>Hemiplegia, neglect, headache</td>
<td>59/m</td>
<td>59</td>
<td>Narrowing of both intradural ICA ending at the intracranial bifurcation (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
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<tr>
<td>34</td>
<td>Global aphasia</td>
<td>65/m</td>
<td>110</td>
<td>Bilateral supraclinoid portions</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td>35</td>
<td>n.a.</td>
<td>72/f</td>
<td>98</td>
<td>Bilateral carotid siphon stenosis (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Sonography, TAB</td>
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<tr>
<td>36</td>
<td>Self-limited upper limb weakness, facial droop, no headache</td>
<td>59/f</td>
<td>97</td>
<td>Stenosis in the ophthalmic segment of the left ICA (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
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<tr>
<td></td>
<td>Vision loss, quadrantanopia, headache</td>
<td>72/f</td>
<td>50</td>
<td>Bilateral carotid siphon stenosis (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td></td>
<td>Vertigo, nausea, sweating, headache</td>
<td>69/m</td>
<td>21</td>
<td></td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>ACR criteria</td>
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<td>Ref.</td>
<td>Neurological symptoms</td>
<td>Age [y]</td>
<td>Sex [m/f]</td>
<td>ESR [mm/h]</td>
<td>Angiography/Autopsy findings</td>
<td>Extracranial involvement</td>
<td>Intracranial involvement</td>
<td>Bilateral involvement</td>
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<tr>
<td></td>
<td>Gait instability, poor limb coordination, headache</td>
<td>73</td>
<td>m</td>
<td>68</td>
<td>Multiple stenoses in the intracranial left ICA (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Narrowing of the cavernous segments of both internal carotid arteries (angiography)</td>
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<td>6</td>
<td></td>
<td>n.a.</td>
<td>n.a</td>
<td>n.a</td>
<td>Extradural stenosis &gt;60% (uni- or bilateral not mentioned)</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
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<td></td>
<td></td>
<td>n.a</td>
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<td></td>
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<tr>
<td>37</td>
<td>Headache, hemiparesis, aphasia, apraxia</td>
<td>75</td>
<td>m</td>
<td>74</td>
<td>Left supracholinoid segment stenosis</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td></td>
<td>Headache, hemiparesis, dysarthria</td>
<td>70</td>
<td>f</td>
<td>108</td>
<td>Bilateral supracholinoid and petrous segment stenosis</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<td>25</td>
<td>Vision loss, headache</td>
<td>66</td>
<td>m</td>
<td>14</td>
<td>Bilateral stenosis of petrous and cavernous segments (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<td>24</td>
<td>Frontal lobe syndrome, gait ataxia, headache</td>
<td>61</td>
<td>f</td>
<td>unknown</td>
<td>Circumferential arterial wall thickening of carotid siphons (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>38</td>
<td>Blindness, hemiparesis, ataxia, headache</td>
<td>67</td>
<td>f</td>
<td>99</td>
<td>Bilateral intracranial stenosis of cavernous and paracnsion segments</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td>39</td>
<td>Episodic double vision and visual blurriness, headache</td>
<td>59</td>
<td>m</td>
<td>50</td>
<td>Bilateral stenosis of the carotid siphons (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td>40</td>
<td>Transient aphasia, headache</td>
<td>69</td>
<td>m</td>
<td>106</td>
<td>Left-sided stenosis of the cervical segment and multifocal stenosis of the carotid siphons and cavernous segment (angiography)</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
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<td>41</td>
<td>Transient palsy and dysphasia, scalp tenderness, no headache</td>
<td>69</td>
<td>f</td>
<td>86</td>
<td>Bilateral stenosis of the carotid siphons (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td>42</td>
<td></td>
<td>75</td>
<td>f</td>
<td>unknown</td>
<td>Obstruction of both internal carotid arteries</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Ref.</td>
<td>Neurological symptoms</td>
<td>Age [y]</td>
<td>ESR [mm/h]</td>
<td>Angiography/ Autopsy findings</td>
<td>Extracranial involvement</td>
<td>Intracranial involvement</td>
<td>Bilateral involvement</td>
<td>Diagnosis confirmed by</td>
</tr>
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<tr>
<td>43</td>
<td>Hemiparesis, tenderness of head, neck and scrotum, headache</td>
<td>61</td>
<td>129</td>
<td>Bilateral stenosis of the carotid siphons (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Giant cells in biopsy of neck and occipital arteries</td>
</tr>
<tr>
<td>44</td>
<td>Ischemic optic neuropathy, headache</td>
<td>60</td>
<td>64</td>
<td>Bilateral carotid siphon arteritis (angiography)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
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<tr>
<td>12</td>
<td>Diplopia, gait disturbance, Horner’s syndrome, hemiparesis, headache</td>
<td>60</td>
<td>43</td>
<td>Bilateral ICA-stenosis of the full length with maximum in siphons, signs of inflammation, and giant cells found in both ICA (autopsy)</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Giant cells in ICA (autopsy)</td>
</tr>
<tr>
<td>1</td>
<td>Brachiofacial palsy, no headache</td>
<td>65</td>
<td>67</td>
<td>Proximal bilateral occlusion (angiography), proliferation of intima, and giant cells in both cavernous segments—(autopsy)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Giant cells in ICA (autopsy)</td>
</tr>
<tr>
<td>23</td>
<td>Palsy and ataxia, headache</td>
<td>74</td>
<td>60</td>
<td>Mild involvement of both carotid sinuses (autopsy)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Blindness, dysphasia, hemiparesis, headache</td>
<td>80</td>
<td>80</td>
<td>Left siphon occlusion, left cervical part inflammation, and right siphon inflammation without stenosis (autopsy)</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Vertigo, blindness, headache</td>
<td>79</td>
<td>58</td>
<td>Mild bilateral siphon inflammation, right-sided cervical course mild inflammation (autopsy)</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td></td>
<td>Lateral medullary syndrome, headache</td>
<td>75</td>
<td>47</td>
<td>Stenosis of both cavernous segments (autopsy)</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>TAB</td>
</tr>
<tr>
<td>45</td>
<td>Ischemic optic neuropathy, hemiparesis, headache</td>
<td>61</td>
<td>45</td>
<td>Long stenotic area in the intracranial part of the left ICA (angiography); GCA in both STA, ICA, ECA, and basilar artery (autopsy)</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>TAB</td>
</tr>
</tbody>
</table>
Discussion

We found that internal carotid artery involvement in GCA with ischemic strokes follows a characteristic pattern with bilateral mostly symmetrical distal ICA stenosis or occlusion (typically cavernous and clinoid segments).

To our knowledge this is the first systematic review examining case series and case reports about GCA patients with ischemic stroke and ICA stenosis/occlusion. This bilateral distal internal carotid artery involvement pattern was mentioned before to be a possible manifestation in GCA patients but not yet suggested as a strong diagnostic hint toward GCA which should lead to further investigation in acute stroke patients, e.g., TAB, sonography, PET-CT, or MR angiography to support clinical suspicion.

It has been reported before that patients with large-vessel-giant-cell-arteritis have less headache, jaw claudication or visual symptoms, are younger than GCA patients with temporal arteritis and their TAB specimens are less likely to yield positive results all of which are essential findings and symptoms for the diagnosis according to current GCA guidelines. To date there is no diagnostic proof but positive biopsy and proof of giant cells which in the case of only ICA involvement is often not feasible. Diagnosing GCA following ACR criteria used to be common years ago but is considered obsolete nowadays. According to the current German guidelines on the management of GCA diagnosis should be made by an experienced interdisciplinary team that considers laboratory and radiological findings as well as suggestive clinical features besides histological proof in its diagnostic work-up. Moreover, we want to emphasize the pivotal role of cranial artery sonography as well as positron emission tomography (PET) in search of large vessel involvement several of the cases we included referred explicitly to the obsolete 1990 ACR criteria to confirm diagnosis. Furthermore, the understanding of symptoms and inflammatory distribution of GCA changed in the course of time so that previous cases might have been misdiagnosed with a higher probability or confused with other types of vasculitis that were less well researched at that time such as PACNS. That accounts for clinical diagnostic precision and for the interpretation of histological specimens especially since giant cells are not a phenomenon exclusively observed in GCA but also (for e.g.,) in PACNS patients. Sensitivity might be compromised by the existence of unusual phenotypes. As a confounding factor cases of proximal ICA stenosis might be solely and coincidingly due to macroangiopathy without giant cell infiltration such as presumably in the cases of de Boysson et al. Plenty of data about vessel involvement in CGA is available but mostly in case reports and smaller series the exact localization of stenosis within the carotid artery is not clarified which lowered the number of included articles. Other patients with ICA stenosis were not included since no ischemic stroke was detected, for e.g., four cases that showed an ICA involvement at the carotid siphons.

We scarcely found comparable report cases in other categories of vasculitis (e.g., TAK, polyarteritis nodosa, Kawasaki disease, ANCA-associated small vessel vasculitis, PACNS,
sarcoidosis, Behcet's disease, and varicella zoster virus vasculopathy). In one case of Behcet's disease a patient had bilateral proximal ICA occlusion. Patients with TAK which is increasingly considered as a spectrum disease along with GCA did not show any internal carotid artery involvement without continuous affection of the common carotid artery in all cases of a recent study.

To a lesser degree ICA stenosis can occur in the short proximal intradural course but rarely involves purely intradural vessels. This “intra-/extradural border” might be caused by different arterial wall features of the intra- and extracranial arteritis. Intradural arteries tend to have much thinner vessel walls with less elastin. Wilkinson and Russell suggested this difference to be the reason of the intradural sparing of GCA since vessel wall elastin is considered to be a major target of inflammation in GCA, however, vessel wall elastin may extend up to 5 mm intradurally which might explain a variable involvement of intradural internal carotid arteries. Several cases reporting intradural wall thickening or stenoses of vertebral arteries in GCA patients can be found. It is noteworthy that using MRI imaging diagnosis of vessel wall inflammation itself can be challenging. According to a recent work of Guggenberger et al VWE caused by prominent vasa vasorum might be confused with large artery inflammation of the proximal intradural ICA and vertebral arteries in elderly subjects.

Fig. 1 A schematic addition of all cases of ICA (internal carotid artery) involvement. A strong focus on the carotid siphons is apparent.

Fig. 2 MRI on the left side diffusion-weighted imaging (DWI), on the right side apparent diffusion coefficient (ADC) showing bilateral infarcts at the time of the initial hospital admission.

The incidence rate of ischemic stroke in patients with GCA has been repeatedly reported ranging between 2.8 and 7%. The precision of determining this incidence rate might be influenced by the fact that also a TAB has a sensitivity between 70 and 90%. Consequently, the incidence rate of GCA-related strokes might have been underestimated in previous studies. Also, the typical features of GCA such as temporal headache and jaw claudication might be caused by different inflamed arteries than those causing ischemic strokes which might add to the underestimation of incidence rate. Interestingly, Cid et al found that a hemoglobin level as a marker of chronic inflammatory response is associated with a lower risk of cerebral ischemic complications. The authors assumed that an intensified neovascularization could be the consequence of inflammation and protective against neural damage in the case of ischemic stroke. Gonzalez-Gay investigated this further and found lower circulating vascular endothelial growth factor in vivo and lower VEGD transcription in patients with severe occlusive disease. Hočevar et al discovered that a higher CRP value increases the risk of ischemic stroke in GCA patients with a similar explanation as the aforementioned authors: “through a local angiogenic function of proinflammatory cytokines.

It has been repeatedly reported that at the time of diagnosis patients are more likely to have an ischemic stroke in the vertebrobasilar region rather than carotid perfused region with an estimated ratio of around 5:1.2 which changes to a significant predominance of ischemic strokes in the anterior circulation months or years after diagnosis. This might represent an approximation of the vertebral/carotid artery stroke ratio to the normal population.

Table 2

<table>
<thead>
<tr>
<th>Cases</th>
<th>New onset headache [%]</th>
<th>Extracranial ICA stenosis [%]</th>
<th>Intracranial ICA stenosis [%]</th>
<th>Bilateral ICA stenosis [%]</th>
<th>Ratio male/female</th>
<th>Patient age</th>
</tr>
</thead>
</table>
Compared with ICA the vertebral artery pattern of involvement seems to be less characteristic and less predictable. According to these findings the typical GCA patients with ischemic stroke were described as “old men” with cardiovascular risk factors and strokes in the vertebrobasilar territory.

Stenosis and VWE might pose a higher risk for ischemic strokes in GCA patients but has not been studied systematically yet. Caselli and Hunder 1988 investigated the occurrence of ischemic strokes in GCA patients during a 3-year study period and found a higher incidence rate of ischemic strokes in patients with carotid disease; however, the latter was defined only by bruits and/or diminished pulses. To our knowledge there are no studies comparing the degree of stenosis (including VWE) with the incidence rate of ischemia. Early treatment seems to be important: a retrospective database study showed a strong focus of GCA-related strokes with a fivefold-increased risk during the active phase of the disease. These findings suggest the necessity of an immediate and effective treatment after diagnosis. Diabetes and hypertension which are known to be independent risk factors for cardiovascular ischemia seem to add to the risk of ischemic strokes during the follow-up of 6 months after diagnosis of GCA.

**Conclusion**

As mentioned by the Chapel Hill Consensus Conference 2012 authors, “if the features of a vasculitis that is confined to one organ indicate that it is a limited expression of one of the
systemic vasculitides, this vasculitis should be considered a limited expression of that category of vasculitis rather than an independent SOV (single organ vasculitis). We want to emphasize that the knowledge of this characteristic involvement pattern of GCA could help to find the right diagnosis in similar patients. This could lead to an earlier immunotherapy and a better outcome of the respective patients.

Conflict of Interest
None declared.

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References


