**Supplementary Methods**

**Search Strategy and Study Selection**

Additionally, we manually searched the references of published articles. Titles and abstracts of identified publications were screened and potentially eligible studies were retrieved for full-text review. We only selected peer-reviewed, clinical studies published in English language, including two or more adult patients (aged ≥ 18 years) diagnosed with PACNS according to Calabrese’s and Mallek’s diagnostic criteria. Consequently, apart from few explicitly mentioned exceptions among the summary of study characteristics, treatment, and outcome, all included cases were either biopsy- or angiogram-confirmed. The term “angiogram-confirmed” refers to any kind of angiography, i.e. digital subtraction, magnetic resonance, and computed tomography angiography. If multiple publications on the same patient cohort were found, we analyzed the most recent or the largest cohort publication to prevent duplicating results from individual patients.9–13 For instance, more than one study by de Boysson and Salvarani are included, however, there are no doublings in our analysis since we strictly separated the respective investigations. If articles described mixed patient populations with different kinds of vasculitis, we only included the PACNS patients in our analysis. We only included studies on patients with PACNS diagnosed according to Calabrese´s and Mallek´s diagnostic criteria. Thus, included patients were either biopsy- or angiogram-confirmed. In 2007, diagnostic criteria of RCVS, a relevant differential diagnosis of PACNS, were published.14 Therefore, data of patients with “angiogram-confirmed PACNS” published before 2007 were excluded since these patients in fact might have a RCVS.14

**Supplementary Figures**

**Supplementary Figure 1.** **PRISMA Flowchart of the Literature Search**. PRISMA, Preferred Reporting Items for Systematic reviews and Meta-Analyses; PACNS, primary angiitis of the central nervous system.

**Supplementary Figure 2. Imaging characteristics separated by histological pattern.** All estimators represent the proportions of abnormal results in relation to the number of performed exams. The size of the estimator is proportional to the size of the cohort in the respective study. The indicator I-squared indicates the heterogeneity of the data. Error bars indicate 95% confidence intervals (CI). Forest Plots are separated by histological pattern: lymphocytic, granulomatous, and necrotizing. MRI, Magnetic resonance imaging; gd, gadolinium; Abnormalities may, e.g., consist of infarction, hemorrhage, or GD-enhancement or other characteristics as defined by the respective author.

**Supplementary Figure 3. Imaging characteristics of autopsy cases.** All estimators represent the proportions of abnormal results in relation to the number of performed exams. The size of the estimator is proportional to the size of the cohort in the respective study. The indicator I-squared indicates the heterogeneity of the data. Error bars indicate 95% confidence intervals (CI). Forest Plots are separated by histological pattern: lymphocytic, and granulomatous. MRI, Magnetic resonance imaging; gd, gadolinium; Abnormalities may, e.g., consist of infarction, hemorrhage, or GD-enhancement or other characteristics as defined by the respective author.

**Supplementary Figure 4. Imaging characteristics of patients with spinal cord involvement.** All estimators represent the proportions of abnormal results in relation to the number of performed exams. The size of the estimator is proportional to the size of the cohort in the respective study. The indicator I-squared indicates the heterogeneity of the data. Error bars indicate 95% confidence intervals (CI). MRI, Magnetic resonance imaging; gd, gadolinium; Abnormalities may, e.g., consist of infarction, hemorrhage, or GD-enhancement or other characteristics as defined by the respective author.

**Supplementary Figure 5. CSF abnormalities in different histological pattern.** All estimators represent the proportions of abnormal results in relation to the number of performed exams. The size of the estimator is proportional to the size of the cohort in the respective study. The indicator I-squared indicates the heterogeneity of the data. Error bars indicate 95% confidence intervals (CI). CSF is considered abnormal when the cell count exceeds 4/µl and/or the protein concentration exceeds 450 mg/l. Forest Plots are separated by histological pattern: lymphocytic, granulomatous, and necrotizing. CSF, Cerebrospinal fluid.

**Supplementary Figure 6. CSF abnormalities in autopsy cases.** All estimators represent the proportions of abnormal results in relation to the number of performed exams. The size of the estimator is proportional to the size of the cohort in the respective study. The indicator I-squared indicates the heterogeneity of the data. Error bars indicate 95% confidence intervals (CI). CSF is considered abnormal when the cell count exceeds 4/µl and/or the protein concentration exceeds 450 mg/l. Forest Plots are separated by histological pattern: lymphocytic, and granulomatous. CSF, Cerebrospinal fluid.

**Supplementary Figure 7. CSF abnormalities in cases with spinal cord involvement.** All estimators represent the proportions of abnormal results in relation to the number of performed exams. The size of the estimator is proportional to the size of the cohort in the respective study. The indicator I-squared indicates the heterogeneity of the data. Error bars indicate 95% confidence intervals (CI). CSF is considered abnormal when the cell count exceeds 4/µl and/or the protein concentration exceeds 450 mg/l. CSF, Cerebrospinal fluid.

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| **Supplementary Table 1. Study characteristics.** |
| **Study** | **Sample size, n** | **Study period** | **Study location** | **Study design** | **Follow-up**  |
| **median, range (months)** |
| Salvarani et al. 202011 | 191 | 1983 - June 2018 | Minnesota, USA | single-center retrospective  | 19, 0-337 |
| Mandel-Brehm et al. 201915 | 7 | NS | Francisco, USA | single-center retrospective  | NS |
| Raghavan et al. 201916 | 128a | 1986-2016  | Ohio, USA | multi-center retrospective  | NS |
| Schuster et al. 201917 | 44 | January 2008 - August 2017 | Hamburg, Germany | single-center retrospective  | 61.2 |
| Sundaram et al. 201918 | 45 | January 2000 - December 2015 | Sree Chitra Tirunal Institute  | single-center, retrospective  | 33, 1-356 |
| Wang et al. 201919 | 15 | March 2015 -December 2017 | Changchun City, China | single-center, retrospective  | 9 |
| De Boysson et al. 20189 | 112 | Since 1996 | France | multi-center retro/prospective  | 57, 12-198 |
| Marrodan et al. 201820 | 15 | May 2007 - December 2017 | Buenos Aires, Argentina | multi-center retrospective  | 8, 1-90 |
| Patel et al. 201821 | 3 | NS | Melbourne, Australia | single-center, retrospective  | NS |
| Rooij et al. 201822 | 2 | NS | Utrecht, The Netherlands | single-center, retrospective  | NS |
| Strunk et al. 201823 | 18 | NS | Münster, Germany | single-center, retrospective  | 25, 4-54 |
| Becker et al. 201724 | 25 | August 2013 - December 2014 | Essen, Germany | retrospective  | NS |
| Niu et al. 201725 | 19b | February 2009 - January 2015 | Suqian, China | single-center, retrospective  | 12, 3-36 |
| Zhu et al. 201726 | 2 | NS | China | NS | 4.3, 1.6-7 |
| Singhal et al. 201627 | 47b | 1998 - 2015 | Boston, USA | single-center retrospective  | NS |
| Torres et al. 201628 | 9 | January 2005 - December 2013 | Philadelphia, USA | single-center, retrospective  | NS |
| Kempster et al. 201629 | 3 | 2003 - 2013 | Melbourne, Australia | single-center retrospective  | NS |
| Salvarani et al. 201512 | 163 | 1983 - 2011 | Minnesota, USA | single-center retrospective  | 12, 0-164.4 |
| Vera-Lastra et al. 201530 | 12 c | 2004 - 2010 | Mexico City, Mexico | single-center retrospective  | 36 |
| De Boysson et al. 201410 | 52 | 1996 - 2012 | France | multi-center retro/prospective  | 35, 2-148 |
| Geri et al 201431 | 18b | January 1992 - April 2009 | Paris, France | single-center retrospective  | 15, 4-53 |
| Kim et al. 201432 | 24a | 1993 - 2003  | Sacramento, USA  | single-center retrospective  | NS |
| Mihm et al. 201433 | 5 | NS | Nordhorn, Germany | multi-center retrospective  | 0.75,0.1-24 |
| Suri et al. 201434 | 8 | 2005-2012 | New Delhi, India | single-center retrospective  | NS |
| Coronel-Restrepo et al. 201335 | 3 | NS | Cali, Colombia | single-center retrospective  | NS |
| Deb et al. 201354 | 18a | August 2008 - June 2010 | Hannover, Germany | single-center retrospective  | NS |
| Oon et al. 201336 | 12 | July 1998 -June 2009 | Melbourne, Australia | single-center retrospective  | 29.9 (mean), 0.25–51  |
| Pagni et al. 201237 | 2 | NS | Monza, Italy | single-center retrospective  | 31, 2-60 |
| Pfefferkorn et al. 201338 | 4 | October 2008 -August 2010 | Munich, Germany | single-center prospective  | 02. Jun |
| Pourmahmoodian et al. 201239 | 2 | NS | Tehran, Iran | single-center retrospective  | NS |
| Kraemer & Berlit 201140 | 21a | 2003 - 2008 | Essen, Germany | single-center retrospective  | 24, 1-226 |
| Pizzanelli et al. 201141 | 8 | September 2004 -November 2009 | Pisa, Italy | single-center retrospective  | 17, 7-62 |
| Myung et al. 201042 | 4 | 1996 - 2007 | Seoul, South Korea | single-center retrospective  | 41, 19-101 |
| White et al. 201043 | 9 | NS | Omaha, USA | single-center retrospective  | 8–411 days  |
| Yin et al. 200944 | 8 | August 1995 -April 2006 | Guangzhou, China | single-center retrospective  | 24 |
| Küker et al. 200845 | 9 | 1998–2006 | Oxford, UK | multi-center retrospective  | 1 year or until death |
| Salvarani et al. 200813 | 2 | NS | Minnesota, USA | single-center retrospective  | 47, 34-60 |
| Josephson et al. 200746 | 6 | January 1993 -April 2002 | San Francisco, USA | single-center retrospective | NS |
| Scolding et al. 200547 | 6 | NS | England and Wales | multi-center retrospective  | 34, 13-130 |
| Volcy et al. 200448 | 5 | March 1991 -July 2001 | Medellín, Colombia | single-center retrospective  | 12 |
| Singh et al. 200349 | 4 | 1997 - 2002 | Tamilnadu, India | single-center retrospective  | 3, 0.75-3 |
| Campi et al. 200150 | 6a | 1993 - 2000 | Milan, Italy | single-center retrospective  | 33, 12-60 |
| Panda et al. 200051 | 3 | NS | Bangalore, India | single-center retrospective  | NS |
| Alrawi et al. 19993 | 22 | 1989–1996 | Michigan, USA | single-center retrospective  | 12 |
| Pomper et al 199952 | 2 | 1986-1997 | Cincinnati, USA | multi-center retrospective | NS |
| Koo and Massey 198853 | 5 | NS | Durham, USA | retrospective | 12, 0.04–18 |

a Data on all patients include cases with angiogram- and biopsy-negative results or cases not clearly fulfilling diagnostic criteria. This applies to the studies by Raghavan et al. (n=48), Kim et al. (n=3), Deb et al (n=5), Kraemer & Berlit (n=2), and Campi et al. (n=3); b In further cases, reported by Geri et al. (n=17), Niu et al. (n=19), and Singhal et al. (n=27), neither brain biopsy nor angiogram could be attributed, although the diagnostic criteria introduced by Calabrese and Mallek were fulfilled. Outcome data reported by Geri et al. comprise information on primary and secondary cerebral vasculitis – data could not be separated; c Minimum age of included patients was 16 years. NS, not specified.

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| **Supplementary Table 2. Summary of therapies for PACNS and outcome.** |  |  |
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| **Study** | **Induction and maintenance therapy** | **Therapy of relapses** | **Outcome and its association with therapy** |
|   |   |  |   |
| Salvarani 202012 | n=191; median follow-up: 19 months. Induction therapy: GC n=184, GC only n=72. IV GC pulse n=86 (1 g/pulse, median: 5 pulses) before oral PDN (median starting dose 60 mg/d, median duration 10 months). PDN + CYC n=90: Daily oral CYC n=62 (median dose 150 mg/d, median duration 7 months), IV monthly pulse CYC n=32 (median dose 1000 mg/month, median duration 4 months). PDN + AZA n=6 (median dose 100 mg/d, median duration 11 months). PDN + MMF (median dose 2000 mg/d) n=13, IVIG n=1, infliximab n=1, RTX n=1, plasma exchange n=2.Maintenance therapy n=35 (19%), median duration 17 months, started at a median time of 6 months after beginning of treatment: After CYC n=31 (34%), after 6 months of PDN n=3, after 8 months of single course of RTX + PDN n=1. AZA (100-200 mg/d) n=19, MMF (2-3 g/d) n=8, MTX (7.5-20 mg/week) n=5. Oral CYC (50 and 125 mg/d for 18 and 4 months) after induction of remission with iv monthly CYC; infliximab (5 mg/kg for 8 months) after oral CYC for 91 months n=1. | Relapses n=58 (30.4%) CYC n=24 (oral n=15, iv n=9), iv GC pulse n=6, MMF n=5, RTX n=3, chlorambucil n=3, etanercept n=1, AZA n=1, plasma exchange n=1. Increased doses of PDN n=14 | Favorable response in 148/177 patients (84%), PDN alone n=58 (83%), PDN + CYC n=69 (81%, oral CYC 85%, iv pulse CYC 73%). mRS 4-6 at last follow-up: PDN alone (28%) PDN + CYC (37%) - no difference in deaths between both groups (about 25%). Relapses: PDN alone (39%), PDN + CYC (28%). PDN + other therapies (mainly AZA and MMF): N=18 (95%) improved within 2 months. No difference between patients treated with oral and iv pulse CYC. No additional effect due to iv pulse GC. MMF treatment associated with significant reduction in disability.Effect of maintenance therapy: More relapses (46% vs. 19%), but less high disability (mRS 4-6; 11% vs. 37%) and deaths (6% vs. 27%). |
| Schuster et al. 201918 | n=44; median follow-up [months (range)]: 61.2Induction therapy: GC pulse (1 g/d for 3-5 d) and/or varying oral GC tapering schemes n=30. Monthly CYC pulses n=33 (750 mg/m² for 6 months; increased dosage of 1000 mg/m² in refractory cases; n=19 with concomitant GCs), RTX n=3 (2 1000 mg infusions separated by 2 weeks, followed by 1000 mg every 6 months), MTX n=2 (0.3 mg/kg/week) or AZA n=1 (2 mg/kg/d). Maintenance therapy within 3 months after induction therapy with CYC or RTX n=36: MTX n=11, MMF 2g/d n=10, AZA n=5 or RTX n=5. 94% of all patients received GC-sparing immunotherapy after induction treatment. After 24 months, 86.4%, and after 48 months, 63.3%.  | Relapses n=26 (59%) | Median baseline mRS: 2. mRS≤2 after 2 years n=31 (79.5%), after 4 years n=19 (63.3%), after 6 years n=13 (65%), 4 deaths (9%).Remission n=30 (68%, n=20 received CYC)), relapses within 6 months after diagnosis under first-line therapy n=14 (n=13 received CYC). Median mRS stable in non-CYC group, improvement by 1 point in CYC group. Six months after initiation of first-line therapy, n=33/42 (79%) had favorable outcome, 25 of whom received CYC. After 2 years, favorable outcome in n=31/39 (80%), n=23 with CYC induction therapy.  |
| Sundaram et al. 201919 | n=45; median follow-up [months (range)]:33 (1-356)Induction therapy: Oral GCs (1mg/kg/d) n=45 + monthly CYC pulses n=10 / oral CYC n=1 / AZA n=3. Median delay from onset of symptoms to initiation of treatment: 103 days (12-1893). Median duration of treatment: 6 months (2-36)  | Relapses n=25 (56%)CYC n=11 | mRS ≤2 at 6 months-follow-up: n=33: GCs n=25 (76%) + CYC n=8 (24%); mRS ≥3 at 6 months-follow-up: n=12: GCs n=9 (75%) + CYC n=3 (25%); 7 deaths (16%) during total follow-up.Initial cognitive impairment, NIHSS ≥5 and abnormal EEG more frequently associated with mRS ≥3 at 6 months-follow-up.Higher initial NIHSS and normal DSA at diagnosis more frequent in relapsing patients. |
| Wang et al. 201920 | n=15Induction therapy: i.v. GCs n=15Maintenance therapy: oral GCs n=15 + CYC n=3 | NS | NS |
| De Boysson et al. 201810 | n=112; median follow-up [months (range)]: 57 (12-198)Induction therapy: GCs n=110 +/- CYC n=89 (median 6 (2-12) pulses over 6 (2-10) months (n=86) or oral CYC (n=3)) or weekly RTX (375 mg/m²) for 4 weeks (n=3).Maintenance therapy: AZA 2mg/kg/d n=41, MMF 2g/d n=4 or MTX 0.3–0.5 mg/kg/week n=7. Maintenance treatment was started at a median time of 4 (3–18) months after initiation of induction therapy for median duration of 24 (6–72) months. | Relapses n=36 (34%): GCs n=36, additional CYC n=9 or RTX n=8. AZA n= 8, RTX n=6 as maintenance (no initial maintenance therapy). AZA continued n=2, RTX as maintenance n=2, switch to MTX n=3 or MMF n=2 (with initial maintenance therapy) | Remission after induction therapy n=106 (95%): GCs only n=14 (Group 1), GCs + immunosuppressant n=40 (Group 2), GCs + immunosuppressant + maintenance therapy n=45 (Group 3). Remission for ≥12 months after diagnosis at last follow-up n=70 (66%). Good outcome (survived with mRS≤2 at last follow-up) n=63 (56%): Group 3 n=36 (80%) > Group 2 n=16 (40%) > Group 1 n=6 (43%); total median mRS at last follow-up: 2. Relapses: Group 2 n=20 (50%) > Group 3 n=9 (20%) > Group 1 n=4 (29%); death n=9 (8%). |
| Marrodan et al. 201821 | n=15; median follow-up [months (range)]: 8 (1-90)Induction therapy: Oral GCs (1 g/d for 5 days) followed by gradual tapering n=15.Maintenance therapy: Oral GCs n=15, CYC n=8, MMF n=2, AZA n=1, RTX n=1. | Relapses n=12 (80%) | Death n=0 |
| Patel et al. 201822 | n=3Induction therapy: Oral GCs (25 mg/d, n=1; 50 mg/d, n=1), i.v. GCs (1000 mg for 5 days, n=1 and 500 mg for 3 days, n=1, both followed by oral GCs), 2 doses of 1 g i.v. RTX n=1, monthly CYC (1g) n=1Maintenance therapy: AZA (2 mg/kg) n=2. | Relapses n=2 (66.7%)Change from i.v. to oral CYC (2mg/kg), followed by 2 doses of RTX (1g) and, temporary, MMF (2g/d) followed by second course of RTX n=1.I.v. GCs(1g) for 3 days + oral CYC (2mg/kg) + transition from AZA to MMF; after second relapse I.v. GCs and I.v. Cyc + 2 doses of RTX (1g) followed by MMF maintenance n=1 | Stable 9 months after treatment with RTX n=1, clinically improved during maintenance therapy with AZA without any relapses n=1, clinically stable after two courses of RTX n=1; death n=0.  |
| Rooij et al. 201823 | n=2GC pulse + AZA n=1 | Relapses n=1 (50%)AZA changed to monthly CYC pulses for 6 months | No new symptoms or radiological abnormalities at last follow-up after switch to CYC pulses n=1 (50%); death n=1 (50%) |
| Strunk et al. 201824 | n=18; median follow-up [months (range)]: 25 (4-54)Induction therapy: i.v. GCs n=3 + CYC n=1, oral GCs + CYC pulses n=1.Maintenance therapy: AZA n=1, MTX n=1. | Relapses n=6 (33%) | Median mRS at last follow-up: 3 (0-6);Remission at last follow-up n=11 (61%), death n=2 (11%) |
| Niu et al. 201726 | n=19b; median follow-up [months (range)]: 12 (3-36)Less severe disease: Oral GCs (1-2 mg/kg/d) n=11; severe condition: i.v. GCs (1g/d for 3 days) + i.v. CYC (10 mg/kg) every 2 weeks for 3 months n=8 | Relapses n=2 (11%) | Stable clinical course n=17 (89%), relapses n=2 (11%), death n=0. |
| Zhu et al. 201727 | n=2; median follow-up [months (range)]: 4.3 (1.6-7)Induction therapy: i.v. GCs (1 g/d for 3 days) n=2Maintenance therapy: oral GCs (1 mg/kg/d) for 4 weeks + CYC 2 mg/kg/d for 6 months) n=1, oral GCs gradually tapered to 30 mg/d, then maintained for 6 months n=1  | Relapses n=0 | Stable state in spite of mild cognitive impairment n=2 (100%), death n=0. |
| Singhal et al. 201628 | n=47 b | NS | mRS score at discharge: 0–1 (n=21, 45%); 2–3 (n=15, 32%), 4–5 (n=11, 23%); death n=0. |
| Vera-Lastra et al. 201531 | n=12c (minimum age of included patients was 16 years); median follow-up [months (range)]: 36Induction therapy: i.v. GC (1 g per day for 3 days) + 1 pulse of i.v. CYC (750–1000 mg/m2) monthly during 6 months + 20–30 mg/d of oral GCs (with a tapering to 10 mg/day) n=12.Maintenance treatment: Oral GCs (30 mgtapering to 10 mg/day) during at least 12 months or addition of AZA (50–100 mg/d). Three patients with severe disease were given 12 months i.v. CYC. | Relapses n=1 (8%) | Death n=0 |
| Geri et al 201432 | n=18b; median follow-up [months (range)]: 15 (4-53)GCs (i.v. pulses in, approximately, one-third of the cases), i.v. CYC, median number of pulses 6. Median duration of treatment 25 (11–66) months  | Relapses n=8 (44%) | Improvement of more than 2 points on mRS in 14 cases (45%); median initial and final mRS scores: 2 (1–4) and 0 (0–3); death n=0. |
| Coronel-Restrepo et al. 201336 | n=3Induction therapy: I.v. GCs (1 g/d for 3 days) n= 3 + oral CYC (100 mg/d) n=1 or i.v. CYC (1 g monthly per 6 doses, n=1 or 1 g single dose, n=1), followed by oral GCs (1 mg/kg/d, n=1 or 50 mg/d, consequently tapered n=1) | Relapses n=1 (33%)Initially, i.v. GCs (2 g/d per 3 days); due to further deterioration RTX (2x1g within 2 weeks) | Clinical recovery n=2, death after relapse n=1 (33%). |
| Oon et al. 201337 | n=12; mean follow-up [months (range)]: 29.9 (0.25-51)Induction therapy: GCs alone n=5, GCs + CYC n=7 (i.v. n=6 (800 to 1000 mg CYC monthly for 6 months, oral n=1, 100 mg/d). GC schemes: i.v. MP n=9, 1 g/d for 3 days. Dexamethasone n=1, oral PDN n=2. Maintenance therapy: AZA + PDN n=3, PDN alone n=3, CYC + oral PDN n=1 | NS | Median mRS at last review: mRS 0–1: n=9 (75%), ≥2: n=3 (25%); mRS=0 at last follow-up and GCs alone as induction therapy n=4; Improvement in symptoms following treatment n=9; death n=1 (8.3%). |
| Pagni et al. 201238 | n=2; median follow-up [months (range)]: 31 (2-60)I.v. GCs (initially 1 g/d, then 2 g/d) n=1 | Relapses n=1 (50%) | Complete remission 5 years after onset of symptoms n=1 (50%), death after disease course of 2 months despite treatment with GCs n=1 (50%). |
| Pfefferkorn et al. 201339 | n=4GCs n=4 + CYC n=3 | Relapses n=0 | Stable clinical course at last follow-up n=3 (75%), death n=1 (25%) |
| Pourmahmoodian et al. 201240 | n=2GCs n=2 | Relapses n=1 (50%)CYC added | Partial recovery at last follow-up n=2 (100%), death n=0. |
| Kraemer & Berlit 201141 | n=21a; median follow-up [months (range)]: 24 (1-226)GCs (i.v. n=9, oral n=14), i.v./oral CYC n=7, AZA n=5, MTX n=4 or MMF n=4 | NS | Median mRS at last follow-up: 2 (range: 1–6); response to therapy n=13 (72%). No significant difference between biopsy and angiogram confirmed diagnostic groups; death n=3 (14%). |
| Pizzanelli et al. 201142 | n=8; median follow-up [months (range)]: 23.8 (7-62)Severe onset (n=4): i.v. MP 1 g/d for 3 days + i.v. CYC monthly up to a total dose of 5–9 g, after CYC MTX i.m. 10–15 mg once a week Less aggressive disease (n=4): i.m. or i.v. GC dose 125–150 mg/d for 3 days, followed by rapid tapering | Relapses n=1 (13%) (severe onset), Infliximab | Less aggressive disease: n=4 (100%) responded to GCs.  |
| Myung et al. 201043 | n=4; median follow-up [months (range)]: 41 (19-101)Oral prednisolone alone n=1, CYC n=2 | NS | Clinical improvement (n=4) with 1 patient suffering from new symptoms during follow-up; death n=0. |
| Yin et al. 200945 | n=8; median follow-up [months (range)]: 24Oral CYC n=3, 100 mg/d for 8–10 months, i.v. MP n=4 1000 mg/d for 3–5 days or i.v. dexamethasone 20 mg/d for 10 days n=1, then oral PDN alone 80 mg/d for 3–6 months, then tapered  | Relapses n=1 (13%) | Good response to GCs and CYC without relapses; residual neurological deficits: n=6 (75%). |
| Scolding et al. 200548 | n=15d; mean follow-up [months (range)]: 58 (18-130)GCs alone (PDN, e.g. 60 mg/d, then tapered or dexamethasone, e.g. 4 mg/d i.v.) n=8, GCs + CYC n=4, GCs + AZA and GCs + CYC + AZA in n=1 patient respectively  | NS | N=6 received GCs and/or further immunosuppression with no clear pattern of response to treatment; death n=7 (47%). |
| Volcy et al. 200449 | n=5; median follow-up [months (range)]: 12GCs only n=5 for 3 to 6 months, then tapered  | Relapses n=0 | At 1-year follow-up: Good outcome without relapses (n=5), mild neurological sequelae (n=2), residual seizures (n=1); death n=0. |
| Singh et al. 200350 | n=4; median follow-up [months (range)]: 3 (0.75-3)Steroids, CYC, AZA and IVIG | Relapses n=0 | Significant improvement clinically and radiologically n=4; death n=0. |
| Campi et al. 200151 | n=6a; median follow-up [months (range)]: 33 (12-60)PDN 60 mg/d + CYC 100 mg/d for 6 weeks, followed by PDN tapered over 6 months n=6 | NS | Clinical improvement: n=2 (33%); stable course of disease: n= 3 (50%); moderately worse: n=1 (17%). Significant decrease in the number and size of enhancing and non-enhancing abnormalities. |
| Panda et al. 200052  | n=3 | Relapses n=1 (33%) | mRS at last follow-up: 6, i.e. death n=3 (100%). |
| Alrawi et al. 19993 | n=22; median follow-up [months (range)]: 12GCs alone, GCs + CYC, other | NS | Despite treatment, only n=10 (46%) showed some recovery.No clear benefit of the addition of CYC to treatment with GCs; death n=2 of 9 patients with 1-year-follow-up (22%). |
| Koo and Massey 198854 | n=5; median follow-up [months (range)]: 12 (0.04-18)PDN alone n=2, 100 mg/d, PDN (80 mg/d) + CYC (2 mg/kg) n=2.  | Relapses n=2 (40%), n=1 while off medication but stabilized withreinstitution of treatment (GCs + CYC)  | Satisfactory treatment response in n=3/4 treated patients; death n=2 (40%). |
|  |  |  |  |

a Data on all patients include cases with angiogram- and biopsy-negative results or cases not clearly fulfilling diagnostic criteria. This applies to the studies by Kraemer & Berlit (n=2) and Campi et al. (n=3); b In further cases, reported by Geri et al. (n=17), Niu et al. (n=19), and Singhal et al. (n=27), neither brain biopsy nor angiogram could be attributed, although the diagnostic criteria introduced by Calabrese and Mallek were fulfilled. Outcome data reported by Geri et al. comprise information on primary and secondary cerebral vasculitis – data could not be separated; c Minimum age of included patients was 16 years. d, Cohort includes nine patients with Amyloid-beta related angiitis (ABRA), data could not be separated. AZA, azathioprine; CYC, cyclophosphamide; GC, glucocorticoids; i.m., intramuscular; i.v., intravenous; ; IVIG, intravenous immunoglobulins; MMF, mycophenolate mofetil; MP, methylprednisone; MRI, magnetic resonance imaging; mRS, modified Rankin Scale; MTX, methotrexate; MVD, medium vessel disease; NS, not specified; PACNS, primary angiitis of the central nervous system; PDN, prednisone; RTX, rituximab; SVD, small vessel disease.