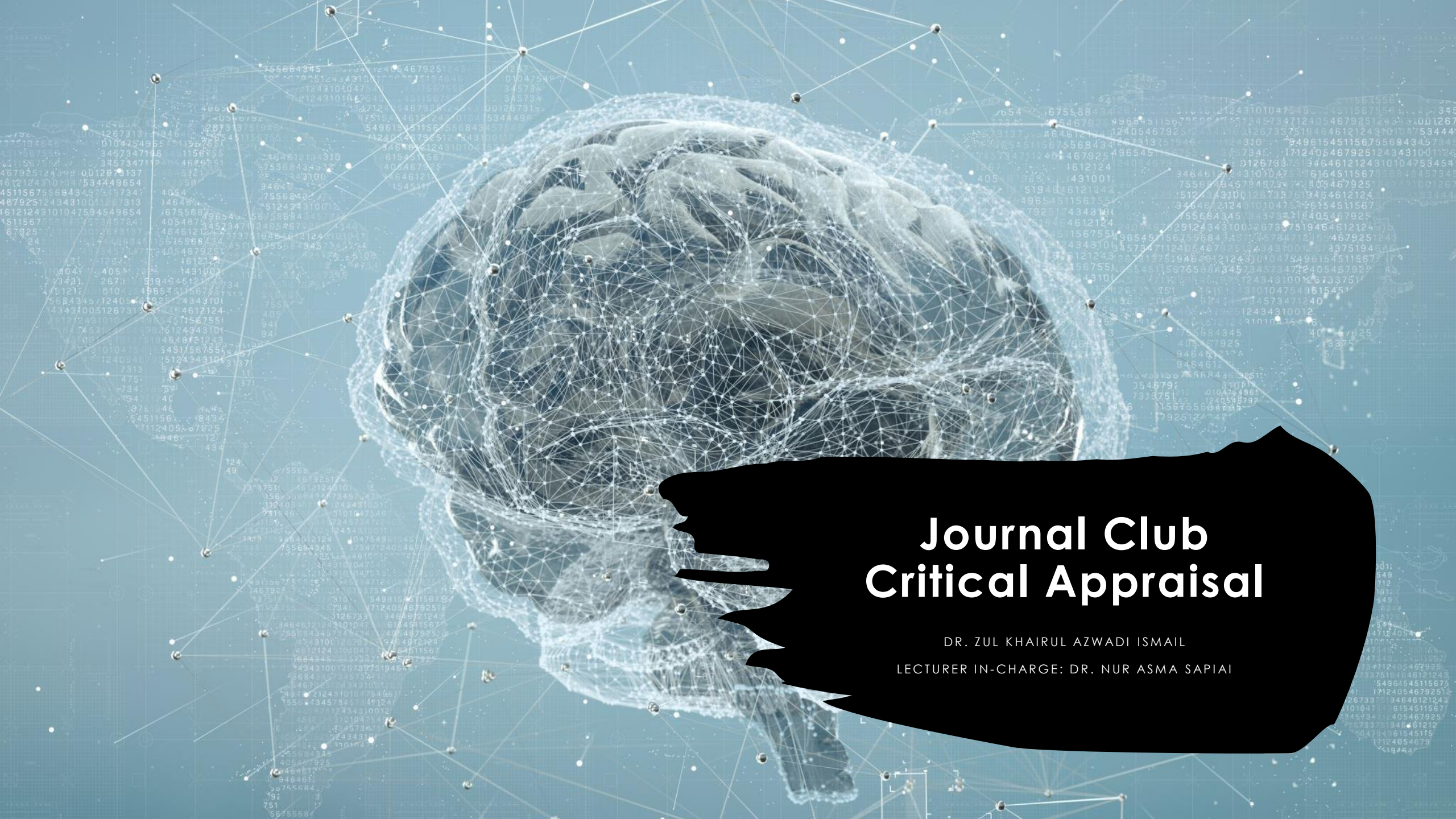

Journal Club Critical Appraisal

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Overview

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ORIGINAL COMMUNICATION

Diagnosis and follow-up evaluation of central nervous system vasculitis: an evaluation of vessel-wall MRI findings

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Single centre study

Abstract

Abstract

Objective To approach the clinical value of MRI with vessel wall imaging (VWI) in patients with central nervous system vasculitis (CNSV), we analyzed patterns of VWI findings both at the time of initial presentation and during follow-up.

Methods Stenoocclusive lesions, vessel-wall contrast enhancement (VW-CE) and diffusion-restricted lesions were analyzed in patients with a diagnosis of CNSV. On available VWI follow-up, progression, regression or stability of VW-CE were evaluated and correlated with the clinical status.

Results Of the 45 patients included, 28 exhibited stenoses visible on MR angiography (MRA-positive) while 17 had no stenosis (MRA-negative). VW-CE was found in 2/17 MRA-negative and all MRA-positive patients ($p<0.05$). 79.1% (53/67) of stenoses showed VW-CE. VW-CE was concentric in 88.3% and eccentric in 11.7% of cases. Diffusion-restricted lesions were found more frequently in relation to stenoses with VW-CE than without VW-CE ($p<0.05$). 48 VW-CE lesions in 23 patients were followed over a median time of 239.5 days. 13 VW-CE lesions (27.1%) resolved completely, 14 (29.2%) showed partial regression, 17 (35.4%) remained stable and 4 (8.3%) progressed. 22/23 patients received immunosuppressive therapy for the duration of follow-up. Patients with stable or progressive VW-CE were more likely ($p<0.05$) to have a relapse (14/30 cases) than patients with partial or complete regression of VW-CE (5/25 cases).

Conclusion Concentric VW-CE is a common finding in medium/large-sized vessel CNSV. VW-CE might represent active inflammation in certain situations. However, follow-up VWI findings proved ambiguous as persisting VW-CE despite immunosuppressive therapy and clinical remission was a frequent finding.

Keywords Cerebral vasculitis · Stroke · Vessel wall imaging · MRI · Follow-up

- Concise
- Emphasize important findings
- Clear conclusion
- Data collection time period NOT mentioned
- Study design (retrospective) NOT stated
- Clinical and histopathological factors as part of diagnosis NOT mentioned

Introduction

- Brief introduction of vasculitis and current methods of diagnosis.
- Concise description on basics of VWI.
- Knowledge/information gap written.
- Study aim stated

Introduction

Central nervous system (CNS) vasculitis is a rare disease characterized by different etiologies, heterogeneous findings and a lack of definite diagnostic markers. Thus it poses great challenges regarding both diagnosis and treatment [1–6].

Since therapy usually consists of long-term immunosuppression with potential major adverse effects [5, 6], it is essential to establish a definite diagnosis and to evaluate the treatment response carefully. Along with clinical and laboratory findings, imaging is crucial in the work-up of CNS vasculitis [7]. However, findings of both digital subtraction angiography (DSA) and conventional magnetic resonance imaging (MRI) including magnetic resonance angiography (MRA) are unspecific regarding CNS vasculitis [8–10]. While evidence of systemic vasculitis or CNS infection can help establish the diagnosis of CNS vasculitis in some cases, brain biopsy is considered mandatory to prove primary angiitis of the central nervous system (PACNS) [5, 7, 11, 12]. Yet even biopsy has a limited sensitivity with a relevant rate of false negative results, particularly when only medium- and/or large-sized vessels are affected [13–17].

Therefore, advances regarding diagnostic tests for CNS vasculitis are required. MRI with dedicated vessel wall imaging has been proposed in this respect [18–20]. Vessel wall imaging uses different techniques to suppress the signal of intraluminal blood ("black blood imaging"), thus allowing evaluation of the vessel wall and possibly the detection of inflammatory changes within the vessel wall [19, 21]. Vessel wall contrast enhancement has been reported as a potential sign of CNS vasculitis as early as 2008 [22]. Vessel wall contrast enhancement in patients with CNS vasculitis is presumed to be caused by hyperpermeability of the endothelium and/or by neovascularization, resulting in leakage of contrast into the arterial wall either from the lumen of the main vessel or from vasa vasorum [19]. Vessel wall imaging is now used in suspected CNS vasculitis in many neurovascular centers [19]. However, there is still an eminent lack of original research on this subject, which is certainly due to the rarity of the disease. To date, there are only a few case reports and series evaluating vessel wall imaging in CNS vasculitis, with 29 patients being the largest reported CNS vasculitis group to our knowledge [23]. Even less data is available concerning follow-up MRI with vessel wall imaging in CNS vasculitis patients. According to our literature research, the largest study groups in which follow-up vessel wall imaging results were specifically reported comprise only six patients [24, 25]. Thus the role of vessel wall imaging both regarding the diagnosis of CNS vasculitis and monitoring disease activity, particularly in response to immunosuppression, remains largely unproven.

It is for these reasons that we retrospectively evaluated clinical and radiological data of patients with CNS vasculitis treated at our institution, aiming to contribute data on the pattern of vessel wall imaging findings both at the time of initial presentation and at follow-up.

Methods

Methods

Patients

We searched the electronic medical records of the Department of Neurology of our institution from September 2008 to December 2019 for adult patients (≥ 18 years) with suspected CNS vasculitis. The time span was chosen because dedicated vessel wall MRI has been used at our institution since September 2008.

In a second step, the diagnoses of CNS vasculitis were reviewed. For the purpose of this study, CNS vasculitis was defined as an inflammatory vasculopathy of cerebral arteries, either restricted to the CNS or as part of a systemic disease. clinical, laboratory, imaging and neuropathological data of each patient were evaluated. Relevant clinical and laboratory findings included the clinical presentation, patient age, the presence of CNS inflammation evidenced by cerebrospinal fluid exams, serologic results including parameters for systemic collagenosis and vasculitis, other evidence of systemic disease and cardiovascular risk profile. Available imaging exams [MRI, MRA, DSA, computed tomography (CT), positron-emission tomography—computed tomography (PET-CT)] were assessed for the presence, distribution and age of ischemic or hemorrhagic brain lesions, cerebral parenchymal or meningeal contrast enhancement, irregularities, stenoses or occlusions of intracranial arteries and signs of systemic vasculitis. The available neuropathological reports on brain and/or meningeal biopsies were reviewed.

- Patient selection and data collection systematically described.
- Complete description of diagnostic parameters collected.
- Specific serologic markers not stated.

Patients were included for further analysis if a diagnosis of definite or probable CNS vasculitis could thus be established. Regarding PACNS, diagnoses were made according to the work-flow suggested by Berlit and Krämer [3]. This work-flow was developed with regard to the diagnostic criteria of PACNS developed by Birnbaum and Hellmann in their 2009 revision [7] of the Calabrese and Mallek criteria [11] (for details see Fig. 1). Patients were categorized according to the affected vessel size as proposed by Küker [26]: DSA-negative patients were classified as having small-vessel CNS vasculitis while patients with stenoses visualized on DSA and/or MRI were classified as having large- and/or medium-sized vessel CNS vasculitis. The large/medium vessel CNS vasculitis group was further subdivided in patients with pathologic findings (luminal irregularities, stenoses, occlusions) visible on MR angiography ("MRA-positive") and patients with luminal abnormalities only depicted by DSA ("MRA-negative").

Subsequently a search of the identified patients in the local Picture Archiving and Communication System (PACS) was performed. Patients who had at least one MRI scan including dedicated vessel wall imaging were included in the study.

- Confusing description on diagnostic criteria for PACNS
  - Berlit&Kramer or Kuker?
  - If Kuker criteria used – why use flow chart of Berlit& Kramer?

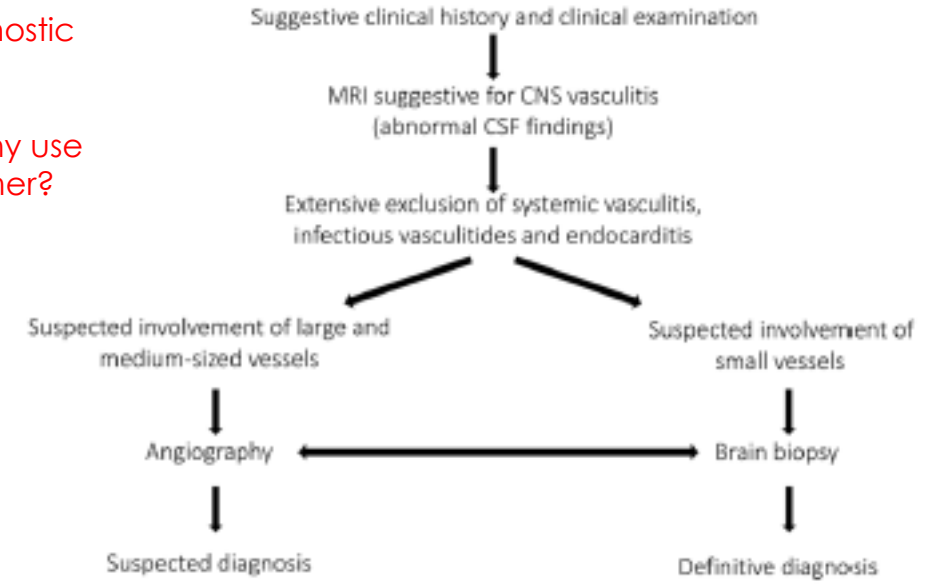


Fig. 1 Flow chart on the diagnostic work-up for PACNS (adapted from Berlit and Kraemer [3]; Birnbaum and Hellmann [7])

MRI protocol

MRI protocol

89% of the analyzed MRI scans were acquired on a 3 Tesla Signa Excite scanner (GE Healthcare, Milwaukee, WI, USA), 8% on a 3 Tesla Magnetom Verio scanner (Siemens Healthineers, Erlangen, Germany) and 3% on a 1.5 Tesla Magnetom Aera scanner (Siemens Healthineers, Erlangen, Germany). The 1.5 Tesla scanner was used for one patient with a cardiac pacemaker not approved for 3 Tesla.

Vessel wall imaging was performed using a fat- and blood-suppressed 2D double inversion recovery spin-echo T1-weighted sequence pre- and post contrast. The sequences were acquired in axial and coronary planes with a slice thickness of 2 mm. Depending on the scanner, the in-plane resolution was 0.5×0.5 mm (Signa), 0.4×0.4 mm (Verio) or 0.9×0.9 mm (Aera). Eight to 14 slices were positioned to include the most prominent stenoses as visualized by Time-of-Flight-MRA. If there were no obvious stenoses, the sequences were positioned to cover the Circle of Willis.

The imaging protocols further included diffusion-weighted imaging (DWI), fluid-attenuated inversion recovery (FLAIR), T2* or susceptibility-weighted imaging (SWI) and, in most cases, contrast-enhanced MR angiography and a 3D T1-weighted sequence pre- and post contrast.

- Type of machine stated.
- Image acquisition described.

Image analysis

Of each patient, all MRI scans including vessel wall imaging acquired within the first two years after initial presentation to our hospital and the last available MRI scan were evaluated. The timing of the initial vessel wall imaging examinations in respect to the time of presentation to our institution as well as the start of immunosuppressive therapy was documented. Follow-up intervals were categorized as "short-term" (within three months after the first MRI with vessel wall imaging), "mid-term" (3–12 months) and "long-term" (> 12 months).

All stenoocclusive lesions depicted on vessel wall imaging were recorded by correlating vessel wall imaging with MRA sequences. The lesions were graded by visual inspection of MRA as 1 = slight narrowing and irregularity of the lumen (estimated stenosis grade < 30%); 2 = moderate stenosis (30–69%); 3 = high-grade stenosis (70–99%) or 4 = occlusion. For each depicted stenoocclusive lesion, the degree of vessel wall contrast enhancement was documented as either 0 = no enhancement, 1 = moderate enhancement or 2 = strong enhancement as defined and shown by Pfefferkorn et al. [29]. Vessel wall contrast enhancement without stenosis was also recorded. Furthermore, the morphology of contrast enhancement on the initial MRI scan was classified as either concentric or eccentric. This classification was performed as previously described by Obusez et al. [25]: Enhancement was recorded as concentric if it was uniform and involved the entire circumference of the arterial wall and as eccentric if it was nonuniform, mainly on one side of the arterial wall and not involving the entire circumference. DWI was examined and diffusion-restricted lesions signifying acute ischemic infarctions were identified. The diffusion-restricted lesions were recorded according to their location in relation to the stenoses depicted on vessel wall imaging, i.e. whether they were sited in a territory supplied by a stenotic artery. In patients with contrast-enhancing vessel wall lesions, each follow-up MRI scan with vessel wall imaging was compared to the previous exam and regression, stability or progression of each contrast-enhancing vessel wall lesion was reported. Newly occurred contrast-enhancing vessel wall lesions were recorded as progression. Lesions which continued to display complete resolution of vessel wall contrast enhancement were categorized together with regressive contrast-enhancing vessel wall lesions. Diffusion-restricted lesions were also recorded on follow-up.

Vessel wall imaging on initial presentation and follow-up was analyzed by two neuroradiologists separately and blinded to other imaging and clinical findings. Discrepant reading results were resolved in consensus.

- Detailed description on image analysis
- Should further clarify DWI lesion in vessels or parenchyma?

Clinical analysis

The medical records of each patient were evaluated and clinical parameters were documented for each patient at the time of each MRI examination. At baseline, the onset of symptoms (i.e. acute versus subacute), the range of neurological symptoms (i.e. headache, focal neurological deficits, cognitive/behavioral changes, newly occurring symptomatic epilepsy/epileptic seizures) and the National Institute of Health Stroke Scale (NIHSS) Score were documented. At each follow-up, the range of neurological symptoms and the NIHSS were again documented (for details see Table 2, Suppl. 1).

Additionally, on follow-up, the clinical status was recorded as either "stable disease/remission" or "relapse". A relapse was defined according to Salvarani et al. [30] as either a recurrence of or worsening of symptoms of CNS vasculitis and/or evidence of new diffusion-restricted lesions or an increase of ischemic lesions on MRI. A relapse usually was associated with an increase in immunotherapy (see Table 2).

Treatment of CNS vasculitis with steroids and/or other immunosuppressive agents was documented at initial presentation and at the time of each MRI examination.

- Clear statement of symptoms and scoring
- Clear case definition for follow-up cases

Statistical analysis

Categorical variables were analyzed using two-sided Fisher's exact test. To evaluate the course of vessel wall imaging findings in relation to clinical findings, the three possible vessel wall imaging outcomes (progressive, stable or regressive/no vessel wall contrast enhancement) were dichotomized in two different ways and compared to the clinical status of "remission" or "relapse", also using two-sided Fisher's exact test. P values < 0.05 were considered significant. Statistical analyses were performed using SPSS Statistics version 25 (IBM, Armonk, NY, USA).

- Brief description of statistical analysis

Results

Results

Population

45 Patients were included in the study. 27 patients were female and 18 were male. The median age was 58 years (range 19–75 years).

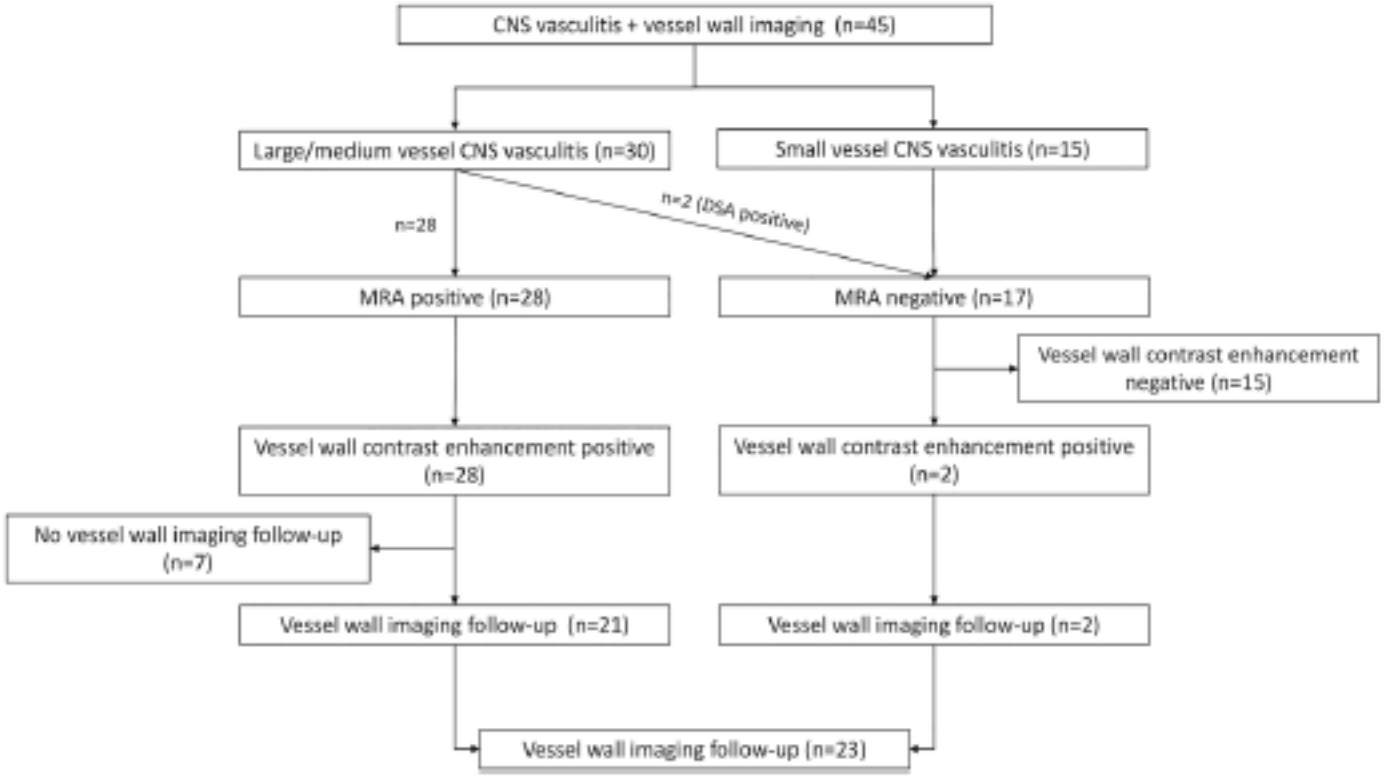
Initially, 15 patients were classified as small-vessel CNS vasculitis and 30 patients as large/medium vessel CNS vasculitis. In 28 of the patients with large/medium vessel CNS vasculitis, stenoses or irregularities of intracranial arteries were visualized on MR angiography ("MRA-positive") while two patients showed abnormalities of medium-sized arteries on DSA only ("MRA-negative"). Consequently, 17 patients were initially categorized as MRA-negative and 28 as MRA-positive. Figure 2 shows the distribution of patients in the different study groups. Table 2 summarizes epidemiological and clinical data, diagnoses and treatment of the patient group.

Forty of 45 patients received their first MRI scan including vessel wall imaging within three months of the initial presentation to our institution. The other five patients were initially examined within four to eight months. Forty-four of the 45 patients were not under immunosuppressive therapy at the time of the first presentation to our hospital. All 23 patients with follow-up evaluations (see below) had their initial vessel wall imaging within four weeks of the initial presentation and none of these patients had received immunosuppression at initial presentation.

- Clear overview of study population
- Not mentioned reason for 7 patients not followed-up
  - Only stated in flowchart.

Results

Fig. 2 Distribution of patients to the different subgroups of the study



Initial presentation

MRA-negative patients

15 of the 17 patients (88.2%) did not show any vessel wall contrast enhancement. Two patients (11.8%) presented with vessel wall contrast enhancement of large arteries without stenosis.

MRA-positive patients

Error! - Not VWI – MRA (page 3 Para 7)

28 patients harbored 67 stenoocclusive lesions examined by vessel wall imaging. The degree of stenosis was graded as "1" in eight cases (11.9%), "2" in 26 cases (38.8%), "3" in 25 cases (37.3%) and "4" in seven cases (10.4%).

53 of the stenoocclusive lesions (79.1%) showed vessel wall contrast enhancement. Vessel wall contrast enhancement without stenosis was found in seven vessel segments in five patients. Any vessel wall contrast enhancement was seen in all patients (100%). Vessel wall contrast enhancement was graded as strong in 51.7% (31/60) and as moderate in 48.3% (29/60) of cases. Vessel wall contrast enhancement was further classified as concentric in 88.3% (53/60) and as eccentric in 11.7% (7/60) of cases.

Group comparison

The presence of any vessel wall contrast enhancement was significantly more frequent in MRA positive vs. MRA-negative patients ($p < 0.0001$) and in large/medium-vessel CNS vasculitis vs. small-vessel CNS vasculitis ($p < 0.0001$).

Correlation of vessel wall imaging and diffusion-weighted imaging

Associated diffusion-restricted lesions were found significantly more often ($p = 0.048$) in stenoses with vessel wall contrast enhancement (18/50, 36.0%) than in stenoses without vessel wall contrast enhancement (1/14, 7.1%). Three stenoses were excluded from this analysis because their association to existing diffusion-restricted lesions could not clearly be determined. Diffusion-restricted lesions unrelated to visible stenoses were found in 13 patients.

No stenosis? Why MRA positive? – See "MRA positive definition" (page 3 Para 1)

- Concise description of main findings at time of diagnosis
- Direct comparison of VWI with MRA & DWI
- Sig. association of VWI-CE with both sequences
- No analysis on HPE as mentioned in Methods.

At DIAGNOSIS

Follow-up

Vessel wall imaging

Twenty-three patients with 48 contrast-enhancing vessel wall lesions had follow-up MRI scans including vessel wall imaging. This comprised seven contrast-enhancing vessel wall lesions that developed during follow-up. 55 MRI scans were analyzed ($n = 1–6$ per patient) and 120 assessments of contrast-enhancing vessel wall lesions were performed overall. The length of follow-up ranged from 7 to 3543 days (Median 239.5 days).

Per contrast-enhancing vessel wall lesion and MRI scan, vessel wall contrast enhancement was graded as progressive in 10/120 cases (8.3%), as stable in 52/120 cases (43.3%) and as regressive/no enhancement in 58/120 cases (48.3%).

Per patient and MRI scan, vessel wall imaging was rated as progressive in 5/55 cases (9.1%), as stable in 25/55 cases (45.5%) and as regressive/no enhancement in 25/55 cases (45.5%).

Short-term follow-up (< 3 months) was available for 16 patients harboring 34 contrast-enhancing vessel wall lesions. Per patient, vessel wall imaging was graded as progressive in one case (6.3%), as stable in seven cases (43.8%) as regressive/no enhancement in 8 cases (50.0%).

Mid-term follow-up (3–12 months) was available for 13 patients harboring 22 contrast-enhancing vessel wall lesions. Per patient, vessel wall imaging was graded as progressive in no case (0%), as stable in three cases (23.1%) and as regressive/no enhancement in ten cases (76.9%).

Long-term follow-up (> 12 months) was available for 9 patients harboring 19 contrast-enhancing vessel wall lesions. Per patient, vessel wall imaging was graded as progressive in three cases (33.3%), as stable in one case (11.1%) and as regressive/no enhancement in five cases (55.6%).

Table 3 Evolution of vessel-wall contrast enhancement on follow-up

Follow-up interval	Complete resolution of VW-CE	Partial regression of VW-CE	Stability of VW-CE	Progression of VW-CE
Entire Follow-up* ($N = 48$)**	13 (27.1%)	14 (29.2%)	17 (35.4%)	4 (8.3%)
Short-term ($N = 34$)**	2 (5.9%)	12 (35.3%)	19 (55.9%)	1 (2.9%)
Mid-term ($N = 22$)**	7 (31.8%)	9 (40.9%)	6 (27.3%)	0 (0%)
Long-term ($N = 21$)**	8 (38.1%)	6 (28.6%)	3 (14.3%)	4 (19.0%)

VW-CE vessel wall contrast enhancement

*Comparison of the initial MRI scan with the last available MRI scan of each patient

**Number of evaluated VW-CE lesions

Diffusion-restricted lesions

Diffusion-restricted lesions were found in nine of 23 patients and in eleven of the 55 follow-up MRI scans. On nine MRI scans, diffusion-restricted lesions were associated with a contrast-enhancing vessel wall lesion while on two scans, diffusion-restricted lesions were unrelated to contrast-enhancing vessel wall lesions (no statistically significant difference).

- Clear categorisation of groups

AT FOLLOW-UP

Stable disease

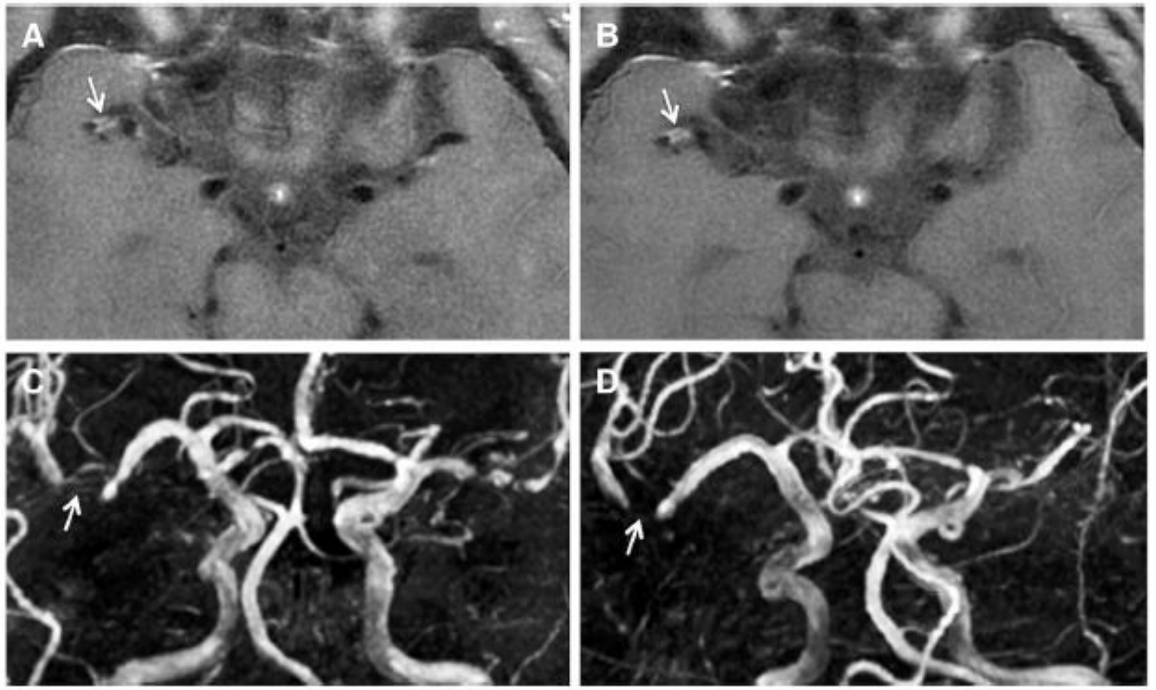


Fig. 3 Stable vessel wall imaging findings on follow-up in a patient with PACNS. Vessel wall contrast enhancement of the right distal M1 segment is seen at initial presentation on vessel wall imaging (**A**), which remains unchanged at two-months follow-up (**B**) despite immunosuppressive therapy. Correlating TOF-MRA findings (**C**, **D**), showing unchanged high-grade stenosis of the affected segment

Disease regression

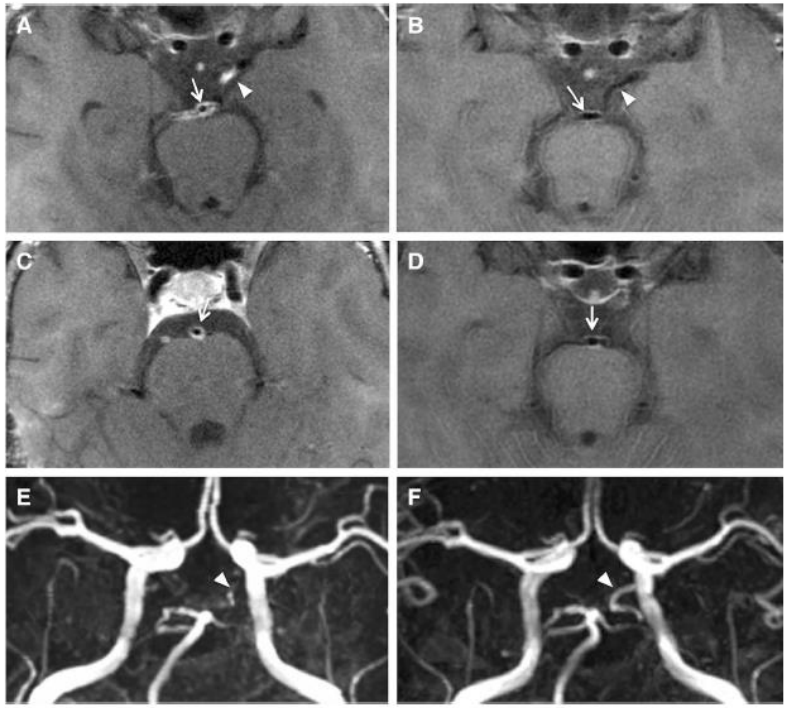


Fig. 4 Regressive vessel wall imaging findings on follow-up in a patient with PACNS. At initial presentation (**A**, **C**), there is marked vessel wall contrast enhancement of the posterior circulation, including the basilar artery (arrow) and left posterior communicating artery (arrowhead). Follow-up vessel wall imaging after ten years (**B**, **D**) shows complete resolution of vessel wall contrast enhancement of the posterior communicating artery and regressive but still persistent vessel wall contrast enhancement of the basilar artery. Correlating TOF-MRA images (**E**, **F**) demonstrate resolution of a high-grade stenosis of the left posterior communicating artery. The findings after ten years are unchanged compared to a six months follow-up scan (not shown). The patient was under immunosuppressive therapy for the whole follow-up period

Progressive disease

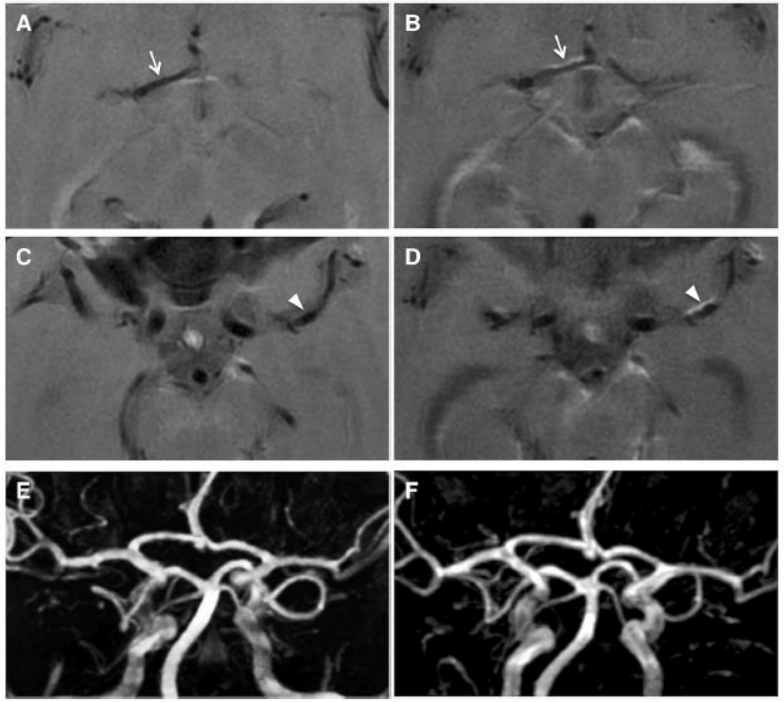


Fig. 5 Progressive vessel wall imaging findings on follow-up in a patient with CNS vasculitis due to cryopyrin-associated periodic syndrome. Follow-up vessel wall imaging performed 34 months after the initial presentation (**B**, **D**) depicts contrast enhancement along the anterior vessel walls of the right A1 segment (arrow) and the left M1 segment (arrowhead), which was not identifiable on the initial MRI scan (**A**, **C**). Perivascular contrast enhancement surrounding the posterior cerebral arteries can be seen on both scans. Correlating TOF-MRA images (**E**, **F**) at both times do not show stenoses of the arteries of the circle of Willis ("MRA-negative"). The patient was under immunosuppressive therapy for the follow-up period

Discussion

Discussion

Regarding the findings at the time of initial presentation of the patients, the results of our study corroborate several common assumptions about vessel wall imaging in patients with CNS vasculitis. Any vessel wall contrast enhancement was reported by Küker et al. in 85.2% and by Thaler et al. in 60.9% of cases with large/medium vessel CNS vasculitis [22, 23]. In our study, more than three quarters of the depicted stenoses showed vessel wall contrast enhancement and any vessel wall contrast enhancement was found in each of the MRA-positive patients. Thus we confirmed that vessel wall contrast enhancement is a frequent finding in patients with large/medium vessel CNS vasculitis. However, it is important to be aware that vessel wall contrast enhancement is not exclusive to vasculitis but can also occur in various other pathologies. Based on an overview of differential diagnoses of PACNS published in a recent review article [31], we compiled a list of different subtypes of central nervous system vasculitis as well as possible differential diagnoses in which vessel wall contrast enhancement has been reported in the literature (see Table 4). From this list it becomes clear that a diagnosis of central nervous system vasculitis cannot be based simply on the presence of vessel wall contrast enhancement. Important potential mimicks of central nervous system vasculitis in which vessel wall contrast enhancement has been shown, at least to a lesser extent, include atherosclerosis, moyamoya disease and reversible cerebral vasoconstriction syndrome (RCVS) [32–36]. However, the morphologic characteristics of vessel wall contrast enhancement could be helpful in distinguishing between distinct vasculopathies. Vasculitis is usually considered to result in concentric wall-thickening and enhancement as opposed to eccentric plaque enhancement in atherosclerosis [18, 19]. Our findings support this assumption, as 90% of vessel wall contrast enhancements were classified as "concentric". This also shows, however, that eccentric vessel wall contrast enhancement can occur in CNS vasculitis in a minority of cases, thus potentially further complicating the differentiation from atherosclerosis. Our results in this regard corroborate the findings of Obusez et al. [25] who reported eccentric wall abnormality in three of twelve CNS vasculitis cases. Further research is needed to define the role of vessel wall imaging in differentiating central nervous system vasculitis from other vasculopathies.

- Comparison with other studies
- Clear description of key distinguishing features of vasculitis vs other dx
- Concentric = vasculitis (90%)
- Eccentric = atherosclerosis
- 10% eccentric = vasculitis
- Stated that further research needed

differences regarding the frequency of vessel wall contrast enhancement, reporting no vessel wall contrast enhancement in six patients with small-vessel PACNS. Our results are in agreement, as vessel wall contrast enhancement was also found significantly less often in small-vessel CNS vasculitis (2 of 17 cases) than in large/medium vessel CNS vasculitis. It is not a surprising result, as the spatial resolution of MRI might be too low to assess very small arteries/arterioles which cannot be evaluated on DSA. Moreover, our vessel wall imaging sequences were placed to depict large- and medium-sized arteries. However, it is an indication that vessel wall imaging usually will not show signs of large-/medium-sized vessel inflammation which is "invisible" on luminal imaging in patients with small-vessel CNS

- Reasons of low sensitivity of VWI to small vessel vasculitis stated.
- Small vessels vasculitis does not involve large or medium vessels

In our group of CNS vasculitis patients, diffusion-restricted lesions were associated with vessel wall contrast enhancement of stenoocclusive lesions at initial presentation. This observation suggests that vessel wall contrast enhancement represents a condition of the vessel wall that predisposes to ischemic stroke. This in turn might indicate that vessel wall contrast enhancement initially represents active inflammation causing prothrombogenic changes in the vessel wall and/or progressive stenosis.

Explanation on association with DWI

[25]. Furthermore, the vessel wall imaging study group of the American Society of Neuroradiology states that, according to their experience, "there may be a discordance between intracranial VW-MR imaging findings and the clinical impression of vasculitis disease activity" [19].

vasculitis patients largely supports this statement. Twenty-two of 23 patients received immunosuppressive therapy for the length of follow-up, which is probably the reason why progressive vessel wall imaging findings were rare. Complete resolution, regression without disappearance and stability of vessel wall contrast enhancement were relatively evenly distributed. Thus, while some patients showed quite obvious treatment response, others exhibited continued enhancement with unclear significance. Even on long-term follow-up, spanning periods of roughly one to ten years, persistence of vessel wall contrast enhancement was a frequent finding (see Fig. 3). Patients with stable or

- Discussion on role of VWI on follow-up
  - Debatable
  - Showed inconclusive findings

Limitations

- Retrospective
- Heterogenous groups of vasculitis – not specific to vasculitis subtypes
- Inclusion of non—inflammatory vasculopathies.
- Non-biopsy proven.
- No control group.

Conclusions

Analyzing a comparably large group of patients, we found that concentric vessel wall contrast enhancement is common in large/medium vessel CNS vasculitis and rare in small-vessel CNS vasculitis. At initial presentation, vessel wall contrast enhancement of a stenosis was associated with an increased probability of ischemic stroke, supporting the assumption that vessel wall contrast enhancement might represent inflammatory activity. This is further substantiated by the fact that patients with stable or progressive vessel wall imaging findings on follow-up evaluations were more likely to have a relapse. However, persisting vessel wall contrast enhancement despite immunosuppressive therapy and clinical remission was also a frequent finding. Overall, follow-up vessel wall imaging findings and their clinical correlation proved ambiguous. Given the rarity of the disease, multicenter pooling of data in large patient registers will be necessary to determine whether vessel wall imaging has value in guiding treatment decisions in patients with CNS vasculitis.

- Main findings stated.
- Require larger pool of patients – potential for diagnostic meta-analysis.

Summary

- Good article with adequate validity.
- Largest number of patients in comparison to other similar studies.
- Clear statement of findings.
- Applicable to our settings with adequate resource and expertise.

HUSM experience

- B780477

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