Review

Combined brain and heart magnetic resonance imaging in systemic vasculitides: fiction or real need?

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ABSTRACT

Systemic vasculitides (SVs) is a group of diseases characterised by inflammation/necrosis of the blood vessel wall in various organs. Simultaneous brain and heart involvement is a cause of increased morbidity/mortality in SV. We aimed to present evidence of concurrent brain/heart involvement in SV and the role of a combined brain/heart magnetic resonance imaging (MRI) in their risk stratification.

Cerebral vasculitis (CV) can be presented as focal deficits, seizures, headache, neuropsychiatric manifestations or cognitive dysfunction and cardiovascular disease (CVD) as myocardial/vascular inflammation, perfusion/function defects and fibrosis. MRI is a non-invasive, non-radiating technique that allows the reliable identification of intraparenchymal brain lesions and the detection of myocardial/vascular inflammation and fibrosis. However, its use in SV is currently hampered by high cost, lack of availability/expertise and lack of awareness among the clinicians. Although there are no clinical data supporting the combined use of brain/heart MRI in asymptomatic SV, it would be called for in cases with clinical suspicion of brain/heart involvement, especially in those at high risk for CVD/stroke such as SLE/APS. Furthermore, it may be of value in SV with multi-organ involvement, cognitive dysfunction or other neuropsychiatric symptoms with concurrent cardiac involvement, presenting as typical or atypical symptoms with normal routine cardiac evaluation, new onset of arrhythmia and/or HF.

Introduction

Systemic vasculitides (SVs) represent a heterogeneous group of diseases characterised by inflammation and necrosis of the blood vessel wall. Patients usually present signs of systemic inflammation (fever, arthralgias, myalgias, weight loss) and concurrent symptoms depending on the organs/systems involved. According to the 2012 revised Chapel Hill Consensus Conference (CHCC), SVs were classified depending on the size of the involved vessel into large, medium and small vessel SVs (Table I). Any delay in recognition and/or treatment confer a worse prognosis in SVs and therefore prompt diagnosis and treatment are of paramount importance (1).

ANCA-associated small vessel vasculitides (AAVs) are the most common SVs in adulthood, with an estimated incidence >15-23/million, usually in patients >65 years of age. The prognosis of untreated granulomatosis with polyangiitis (GPA), an AAV, is worse than the prognosis of most frequent cancers and mortality rate at 1 year is 80%. The introduction of immunosuppressive treatment has dramatically improved survival; however, 50% of survivors experience a relapse. Vasculitis is associated with an increased risk for cardiovascular disease (CVD) morbidity/mortality, primarily involving brain and heart. Patients with AAV have a 2- to 4-fold increased risk of coronary heart disease compared to controls (2, 3). Suppiah *et al*. (4) found that out of the 535 included patients, 14% had at least one CV event within the first 5 years of follow-up. These included 6% CV deaths, 5% non-fatal strokes and 8% non-fatal myocardial infarction. Older age was associated with higher risk for a CV event and c-ANCA positive patients showed a reduced risk for CV events compared to p-ANCA positive (4, 5).

Brain and heart involvement in systemic vasculitides

SVs may involve any organ of the body. However, it is the combination of heart and brain that is most detrimental (4). Cerebral vasculitis (CV) can present as focal deficits, seizures, headaches, neuropsychiatric manifestations or cognitive dysfunction (6). CVD in SV is due to both traditional and non-traditional risk factors and vascular inflammation. According to biopsy studies, small vessel inflammation leads to premature CVD, myocardial ischaemia and HF (7). There are only few reports presenting simultaneous involvement of heart/brain during SVs (8-12). However, combined assessment of the brain/heart has never been proposed as part of the routine diagnostic algorithm for SVs. Our aim in this review is to describe the vasculitic lesions in the brain/heart and to discuss the potential role of a combined brain/heart MRI evaluation in the risk stratification of SVs.

Magnetic resonance imaging for evaluation of cerebral vasculitis

MRI is the most commonly used imaging technique for the evaluation of suspected CV, due to its capability of visualise different pathologies. A standard brain MRI protocol for CV evaluation should include (13, 14):

a) spin-echo T1- and T2-weighted imaging;
b) fluid-attenuated inversion recovery (FLAIR) imaging;
c) diffusion-weighted (DW) imaging;
d) susceptibility-weighted (SW) imaging;
e) time-of-flight (TOF) MR angiography (MRA);
f) contrast-enhanced T1-weighted imaging (using fat sat suppression and flow compensation) and T1-weighted images (3 mm or less) of areas of abnormality on MRA/TOF;
g) Additional MRI sequences, such as contrast-enhanced high-resolution MRA and perfusion MRI may be needed in selected cases.

T2-weighted images are used for the detection of ischaemic lesions and frank infarction. FLAIR images facilitate the diagnosis of lesions within the subarachnoid space and of ischaemic lesions in white matter. Contrast-enhanced T1-weighted images may reveal leptomeningeal enhancement or coexisting intraparenchymal lesions. During the acute stage of cerebral infarction, DW MR imaging may discriminate acute from chronic ischaemic abnormalities. Furthermore, wall thickening and intramural contrast uptake are frequently found in active vasculitis affecting the large brain arteries (14-17). SW MR imaging greatly contributes to the detection of micro-haemorrhagic lesions associated with CV (18).

Perfusion MR imaging plays an important role in the assessment of blood flow in patients with CV. Two perfusion MRI protocols are currently used:

a) dynamic susceptibility contrast-enhanced MRI, based on the evaluation of first pass contrast agent that leads to "physiologic maps" and
b) arterial spin labelling, that relies on the evaluation of "unlabelled" (subtracted from "labeled") MR images (19).

Conventional MRA can detect changes in arteries but has limited resolution (20). Intracranial TOF sequences can detect stenoses with high sensitivity, in addition to MRA. Finally, contrast-enhanced high-resolution MR imaging at 3.0 T can reliably assess thickening and wall enhancement in vascular stenoses (21).

Cardiovascular magnetic resonance imaging for evaluating cardiovascular disease in systemic vasculitides

Cardiovascular magnetic resonance imaging (CMR) has already been used for the evaluation of CVD, because of its excellent reproducibility and the capability of characterising tissues. In SVs with clinically silent CVD, CMR is of great value in the early diagnosis and follow-up (22).

CMR evaluation of SVs includes the use of the following sequences (14):

a) Steady-state free precession imaging (SSFP) for evaluation of biventricular function.
b) T2-W imaging (oedema imaging) for myocardial and vessels disease acuity.
c) Early (EGE) and late (LGE) gadolinium enhanced T1-W imaging for detection of inflammation and fibrosis, respectively.
d) Pre- and post-contrast enhanced evaluation of great vessels for assessment of vessel wall and vessel patency, respectively.
e) T1-, T2-mapping and extracellular volume fraction (ECV) for evaluation of diffuse fibrosis and quantification of myocardial oedema, respectively.

Table I. SV classification according to Chapel Hill.

Large vessel	Medium vessel	Small vessel
Takayasu's vasculitis	Polyarteritis nodosa (PAN)	ANCA associated vasculitides
Giant cell vasculitis	Kawasaki disease	Immune complex small vessel vasculitides

Brain and heart involvement in large-vessel SVs

Takayasu's arteritis (TAK)

Cerebrovascular manifestations of TAK include ischaemic attacks, stroke and/or hypertensive encephalopathy (23). MRA has a sensitivity and specificity of almost 100% for the diagnosis of TAK (24). T2-weighted MRI shows wall thickening and hyperintensity of inflamed vessel walls with 11% of TAK presenting evidence of stroke (25-27) (Fig. 1).

Cardiac complications are due to hypertension, coronary/pulmonary artery inflammation, aortic regurgitation or autoimmune myocarditis (28) and may lead to myocardial ischaemia, myocardial infarction and/or HF (29) (Fig. 2).

Giant cell arteritis (GCA)

In GCA, contrast-enhanced high-resolution MRI reveals wall thickening and enhancement of involved vessels (30-32). Vessel wall oedema can be detected by T2-weighted images and high signal intensity of contrast enhancement that are typical of disease acuity (33).

GCA can potentially affect the aorta and its branches, leading to myocardial infarction (34), stroke (35), aneurysms (33) and peripheral thrombosis (36-39).

Brain and heart involvement in medium vessels SVs

Polyarteritis nodosa (PAN)

PAN usually affects vessels' bifurcations resulting in micro-aneurysms,

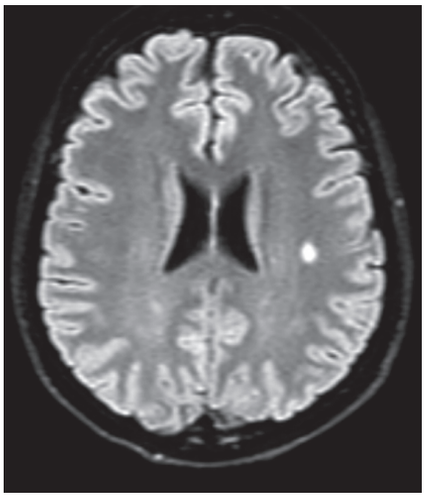

Fig. 1. Asymptomatic cerebral lesions in a patient with TA.

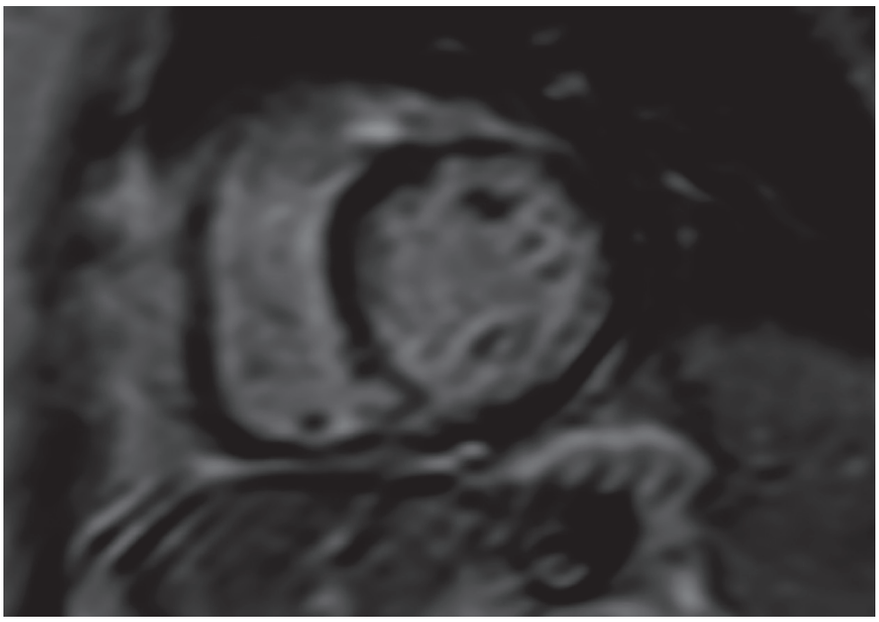

Fig. 2. Subepicardial fibrosis in the inferior wall and supraventricular tachycardia in the same patient.

haemorrhage and/or thrombosis that finally lead to myocardial ischaemia and infarction (40-42). Brain involvement has been reported in up to 20% of PAN. However, since most of the reports were performed before the Chapel Hill criteria, the true incidence is expected to be lower. Central nervous system (CNS) lesions may occur 2–3 years after disease onset (43).

Kawasaki disease (KD)

Up to 30% of KD present with CNS involvement including subdural effusions, cerebral infarctions, atrophy, oedema of the corpus callosum, subcortical lesions and posterior reversible encephalopathy (44-48). Autopsies in KD revealed cerebral vasculitis with endoarteritis and periarteritis (49). Diffuse microhaemorrhages represent a very specific radiological sign of brain vasculitis (50).

The incidence of coronary artery aneurysms (CAAs) in KD was 25% before the introduction of immunoglobulin treatment, but is currently 5–10% (51, 52). However, aneurysms may also occur in other arteries (52-54). Unresolved CAAs develop stenotic or thrombotic lesions leading to myocardial infarction (55).

Brain and heart involvement in small vessels SV

IgA vasculitis (Henoch-Schönlein purpura syndrome)

IgA vasculitis was initially considered a children-exclusive disease, but recently it has also been reported in adults (56). CNS findings are rare and related to hypertensive or uremic encephalopathy (57). Rarely, it can lead to myocardial infarction, due to coronary vasculitis (58).

Microscopic polyangiitis (MPA)

MPA may involve both the nervous system and heart (59, 60). Small-vessel disease involving both white and grey matter is discovered in 37–72% of MPA (59, 60). Coronary ectasia and myocardial infarction have also been described (61).

Granulomatosis with polyangiitis (GPA) (Wegener granulomatosis)

In GPA the CNS is involved in up to 35% of patients (62). MRI of the paranasal sinuses and mastoids are important in the evaluation of cerebral involvement, to rule out continuous extension.

Clinically overt cardiac involvement is rare although coronary artery vasculitis, cardiac arrhythmias, pericarditis, myocarditis, valvulitis and myocardial infarction (MI) have been described (63-70). However, histopathologic studies demonstrated cardiac involvement in 30% of GPA (68).

Eosinophilic granulomatosis with polyangiitis (eGPA) (Churg-Strauss syndrome)

CNS involvement is observed in 6–8% of eGPA (71) and includes cerebral ischaemic or haemorrhagic changes that may lead to confusion, seizures and coma (72) (Fig. 3). Cranial nerve involvement is frequent with the commonest manifestation being ischaemic optic neuropathy. MRI findings vary widely and manifest as macro- or micro-infarctions and haemorrhages (73, 74). ANCA-positive patients were more likely to have mononeuritis, whereas ANCA-negative more likely to have cardiac involvement (75). According to a recent study, endomyocarditis was found in the majority of eGPA with cardiac symptoms and was associated with poor outcome (Fig. 4) (76).

Brain and heart vasculitis of variable-sized SVs

Adamantiades-Behçet's disease (ABD)

In ABD, neurologic disease is diagnosed in 5–30% of patients and is distinguished into 2 types:

a) parenchymal type (80%) involving the brainstem and presenting as hemiparesis, meningoencephalitis, spinal cord and cranial nerve disease;
b) non-parenchymal type (20%), which includes dural sinus thrombosis and arterial occlusion or aneurysms (77, 78).

On T2-weighted images, parenchymal lesions appear as hyperintense areas affecting the brainstem, basal ganglia, periventricular regions, spinal cord and cranial

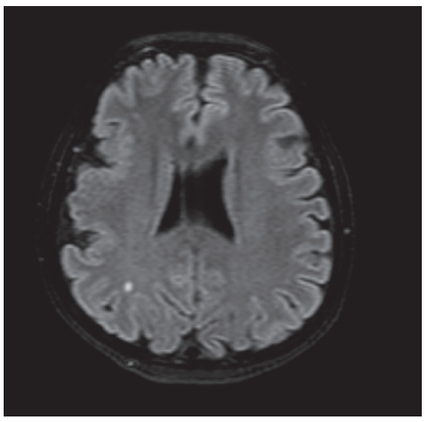

Fig. 3. Asymptomatic cerebral lesions in a patient with CSS.

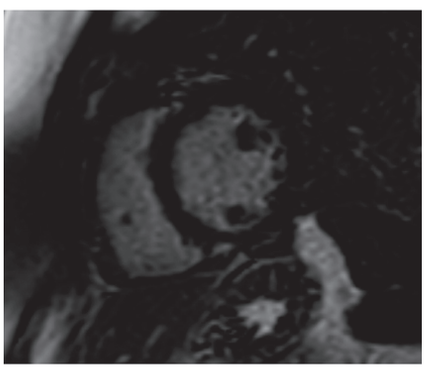

Fig. 4. Subepicardial fibrosis in the intraventricular septum and diffuse subendocardial fibrosis accompaned by pulmonary oedema in the same patient.

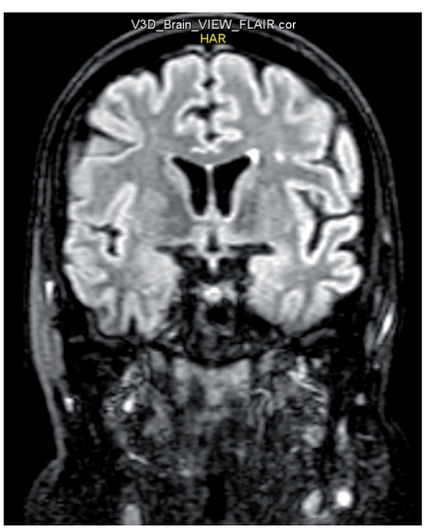

Fig. 5. Asymptomatic cerebral lesions in a patient with SLE.

nerves. Spinal cord lesions are visible on T2-weighted images and enhance after contrast agent injection (78, 79).

Cardiac lesions include pericarditis, endocarditis, intracardiac thrombosis, valvular disease, MI, endomyocardial fibrosis and coronary artery aneurysms. Several cardiac manifestations may coexist in the same patient (80).

Cogan syndrome (CS)

Neurologic symptoms in CS are found in 30% of patients and present with a variety of manifestations ranging from headache and stroke to psychosis and coma. Vasculitis is observed in 12–15% (81). MRI may also show narrowing of the vestibular labyrinth with enhancement in contrast-enhanced T1-W imaging (82).

CS has a mortality rate of approximately 10%. Relevant causes of death include cardiac complications (ruptured aortic aneurysms, myocardial infarction, HF), cerebrovascular and subarachnoid haemorrhage (83).

Brain vasculitis associated with systemic autoimmune disease

Systemic lupus erythematosus (SLE)

Neuropsychiatric SLE (NPSLE) occurs in 14–75% of patients and is associated with a mortality rate of 7–40% (84). Neurologic manifestations including stroke, epilepsy, headache and cognitive dysfunction with white matter involvement are the commonest lesions (60–86%) (Fig. 5). MRA may show stenosis or occlusion of intracranial carotid arteries. Spinal cord myelopathy may coexist with optic neuritis and a long-segment central T2 hyperintensity is the main MRI finding (85-87).

Cardiovascular disease develops in the majority of SLE. The most common findings include pericarditis, myocarditis (usually silent) (Fig. 6) and Libman-Sacks endocarditis, the latter noted in >40% of hearts at autopsy. Severe coronary atherosclerosis leads to myocardial infarction in young adults (88).

Sjögren's syndrome (SS)

CNS disease in SS has been reported in 25–30%. It presents as trigeminal neuropathy, aseptic meningoencephalitis, and uni- or multifocal cerebral disease (89). MRI shows extensive white and grey matter lesions (89, 90).

Primary SS is rarely associated with cardiac involvement. Recently, myocarditis has been described alone or in the context of unresponsive multisystem involvement, with good response to corticosteroid therapy (91).

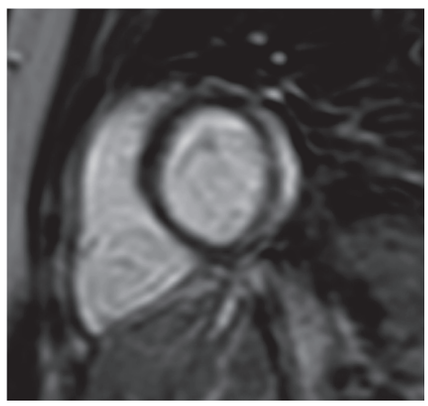

Fig. 6. Evidence of myo-pericarditis in the same patient.

Rheumatoid arthritis (RA)

CNS involvement in RA includes pachymeningitis, dural nodules and, rarely, cerebral vasculitis (92). Rheumatoid pachymeningitis shows increased T2 signal in the subarachnoid space and leptomeningeal contrast enhancement, due to vasculitis. CNS vasculitis is rare and occurs in patients with long-standing active disease (93).

CVD in RA usually occurs a decade earlier than age- and sex-matched controls, and patients with RA are twice more likely to develop MI irrespective of age and traditional CVD risk. RA patients are also at increased risk of atherosclerosis, HF, valvular disease and myopericarditis (94).

Antiphospholipid syndrome (APS)

MRI in APS frequently shows white and grey matter abnormalities, due to small-vessel vasculopathy. DW imaging may show acute infarctions, whereas gradient-echo and SW show microhaemorrhagic areas (95, 96).

Cardiac involvement in APS may present as valvular disease (affecting approximately a third of patients) or less frequently as intracardiac thombosis, pulmonary hypertension, HF, coronary artery or micro-vascular disease with overt or silent clinical presentation. CMR has identified an unexpectedly high prevalence of occult myocardial scarring and endomyocardial fibrosis in APS (97).

Systemic sclerosis (SSc)

MRI is the best non-invasive screening tool for the evaluation of CNS vasculopathy in SSc patients. Reduced brain flow in SSc may finally lead to brain hypoperfusion and cognitive dysfunction (98, 99). Neurological signs and symptoms in localised cutaneous SSc (lcSSc) include epilepsy, headache, focal neurologic deficits as well as neuropsychiatric symptoms (98, 100-107). Brain MRI images show evidence of infarctions in medium-sized arteries and intracerebral haemorrhages (108).

Cardiac involvement in SSc presents with HF, myopericarditis, fatal arrhythmias, pulmonary hypertension, conduction abnormalities and valvular disease, usually with non-specific clinical signs and symptoms. CMR in SSc can reveal oedema, microvascular disease and replacement or diffuse fibrosis (109).

Sarcoidosis (SRC)

In SRC both brain and heart may be affected. Involvement of the CNS occurs in 5–15% of cases. Strictly neurologic forms are seen in fewer than 10% of patients (Fig. 7).

MR findings vary and include white matter lesions on T2W spin echo images mimicking multiple sclerosis, multiple supratentorial and infratentorial brain lesions mimicking metastases, solitary parenchymal mass mimicking high grade astrocytoma and/or solitary non-parenhcymal mass mimicking meningioma. Therefore, these findings are not specific for sarcoidosis and should be considered in the differential diagnosis of other entities presenting with similar findings (110) (Fig. 8).

In heart, CMR can detect inflammation, fibrosis and perfusion defects. Of all cardiac tests, CMR was the most valuable in the diagnosis and prognosis of cardiac SRC (111). Additionally, the presence of myocardial scar indicated by LGE was the best independent predictor of adverse events (112).

MRI patterns can differentiate systemic vasculitis from other common vasculitis affecting the brain/heart

CV is a heterogeneous group of disorders with diverse clinical manifestations that may affect variable sized vessels and be part of a systemic connective tissue disorder. However, their diagnosis becomes particularly challenging if cerebral vessels are affected in isolation or if they constitute the first manifestation of a systemic disease. In this setting, more invasive methods such as digital subtraction angiography (DSA) or even brain biopsy may be included in the diagnostic process. Therefore, there is an emerging need to identify characteristic MRI patterns, which differentiate CV from other common brain pathologic processes, such as reversible vasoconstriction syndrome or intracranial atherosclerosis (113-115).

Imaging findings of cerebral vasculitis are divided in two main categories:

1. Indirect signs reflecting the results of inflammation on vessel morphology or the brain parenchyma such as ischaemic brain lesions or perfusion deficits, intracerebral or subarachnoid haemorrhage and/or vascular stenosis;
2. Direct signs of the pathologic process such as vessel wall thickening with contrast enhancement (116).

Multi-territorial infarcts of different ages are thought to be suggestive of systemic vasculitis, especially if the lesions do not have a typical embolic pattern. High prevalence of haemorrhagic transformation of ischaemic lesions has been recently reported (90). Haemorrhage can be the result either of vessel wall thickening or of ischaemia-reperfusion secondary to vessel stenosis (117).

Leptomeningeal enhancement, especially in association with convexity subarachnoid haemorrhage is highly specific feature of a benign primary angiitis of central nervous system (PACNS) subtype with small leptomeningeal artery vasculitis (118).

MRA can demonstrate multiple stenoses of large and medium sized cerebral vessels. Conventional DSA remains the gold standard in detecting abnormalities especially of smaller vessels. However DSA still lacks sensitivity in demonstrating abnormalities of less than 500μm vessels (119) and given that it is an indirect method, the specificity of this interventional procedure is low. Furthermore, DSA cannot discriminate between underlying causes (120).

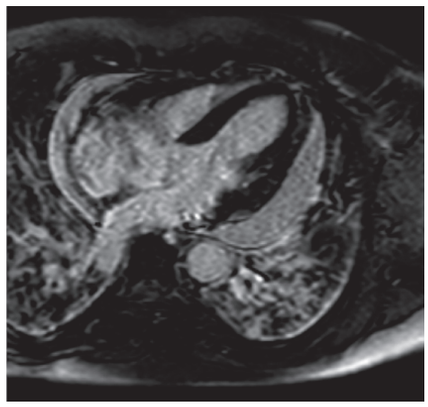

Fig. 7. Asymptomatic cerebral lesions in a patient with sarcoidosis.

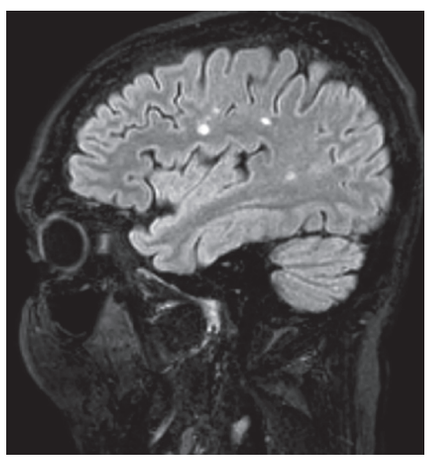

Fig. 8. Subepicardial fibrosis in the lateral wall, due to myocarditis in the same patient.

The main disadvantage of angiographic methods is the inability to differentiate between cerebral vasculitis and other common causes of vascular stenoses, especially atherosclerosis. It is well known that atherosclerosis is relatively rare in young patients without risk factors and has sites of predilection such as arterial bifurcations. Recently, contrast-enhanced high-resolution MR imaging at 3.0 T (21) was proven to be a very useful tool for assessing the vessel wall and differentiating between enhancement patterns of intracranial atherosclerotic plaques and inflammation of other pathology. Atherosclerotic plaques show eccentric irregular wall thickening and gadolinium enhancement of the plaque correlates with plaque instability. In contrast to atherosclerosis, SV produces smooth circumferential concentric wall thicken-

ing with diffuse gadolinium enhancement of the inflamed wall. This wall enhancement is an established imaging sign of vessel wall inflammation (121).

Combined brain and heart MRI in systemic vasculitis: when and why

MRI represents an excellent diagnostic tool, because it is non-invasive, highly reproducible, non-radiating and therefore ideal for the evaluation of chronic diseases, such as SV. However, its use is currently hampered by high costs, limited availability/expertise and lack of awareness among clinicians about its contribution in SV decision making.

To our knowledge, there are no recommendations regarding the potential use of a combined MRI approach in treatment naïve SV patients, except of one paper by our group supporting the role of CMR in both rheumatic and cardiac medication modification in these patients (122). However, until more clinical data become available, a combined brain and heart MRI may be considered for:

a) Patients at high risk for CVD and stroke such as SLE and APS;
b) SV patients with evidence of cardiac involvement. Based on our experience (unpublished data) the majority of SVs with abnormal CMR findings had also concurrent brain lesions;
c) SV patients with cognitive dysfunction and/or other neuro-psychiatric symptoms;
d) SV patients with persistent atypical cardiac symptoms and normal routine non-invasive and/or invasive cardiac examinations;
e) SV patients with new onset arrhythmia and/or HF;
f) SV with multi-organ involvement.

Conclusions

Brain and heart involvement constitutes the major cause of increased morbidity/mortality in SVs. A combined brain-heart MRI evaluation can potentially contribute to better risk stratification in this context. Although no data currently support the use of combined brain-heart MRI in asymptomatic SV patients this may be considered in those at high risk for CVD/stroke (SLE, APS), with neuropsychiatric symptoms, multi-organ and/or cardiac involvement (new onset arrhythmia and/or HF).

Competing interests

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