

Case report

Cardiac amyloidosis presenting with recurrent ischaemic strokes

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SUMMARY

A 72-year-old man presented to our service with sudden onset right-sided weakness, aphasia and gaze palsy with diplopia. CT angiogram demonstrated an acute thrombotic occlusion of the distal basilar artery, a basilar infarct and the patient underwent successful thrombectomy. ECG and telemetry demonstrated slow atrial fibrillation (AF). His transthoracic echocardiogram (TTE) showed a reduced ejection fraction of 25% with global hypo-kinesis, a dilated left ventricle (LV) and LV hypertrophy (LVH). Repeat TTE appeared suspicious for an infiltrative cardiomyopathy with LVH and a speckled appearance to the myocardium. Approximately 10 months later, he suffered another ischaemic stroke post-elective cardioversion for AF while on anticoagulation. Cardiac MRI demonstrated areas of delayed gadolinium enhancement consistent with amyloidosis. Fat pad biopsy was positive for amyloidosis. Our patient has made an excellent recovery from the ischaemic strokes and is being managed in our heart failure clinic.

BACKGROUND

Amyloidoses constitute a group of diseases produced by the deposition of amorphous fibrillar proteins in the extracellular spaces of the body's organs.¹ Cardiac amyloidosis (CA) may be a consequence of any of the systemic amyloidosis. The most commonly encountered systemic amyloidosis that affect the heart are immunoglobulin light-chain (AL) amyloidosis and transthyretin amyloidosis.² The myocardium is affected in almost 50% of AL amyloidosis patients.³⁻⁴ Involvement limited to the heart is rare.⁵ CA exhibits itself with a wide variety of pathological processes, including congestive heart failure, arrhythmia, heart block and cardiomyopathy.⁶

Though not definitive, transthoracic echocardiogram (TTE) and cardiac MRI (CMR) may contribute to additional diagnostic testing and management of the pathology.⁷ Biopsy of the rectum or subcutaneous fat with the use of congo red staining is positive in 80% of patients with systemic amyloidosis.⁸ Endomyocardial biopsy, however, is the gold standard for diagnosis of CA.⁹

Thromboembolic events are a forgotten complication of CA. The Mayo amyloid autopsy study in 2007 showed that intracardiac thrombosis were found in over 50% of AL CA patients.¹⁰ Investigations for patients presenting with cerebrovascular accidents may lead to the discovery of CA incidentally.¹¹ The aim of this case report is to highlight

that amyloidosis can present initially as stroke and to demonstrate that these patients are at a higher risk of thromboembolic events.

CASE PRESENTATION

A 72-year-old man with a background of hypertension, osteoarthritis, vertigo and carpal tunnel syndrome, presented with sudden onset right-sided weakness, aphasia and gaze palsy with diplopia. National Institutes of Health Stroke Scale (NIHSS) was 16. CT of the brain and CT angiogram (CTA) demonstrated an acute thrombotic occlusion of the distal basilar artery. The patient was thrombolysed and immediately transferred to neurosurgery for thrombectomy, which was successful with an NIHSS of 3 day one post-thrombectomy. MRI of the brain post-thrombolysis revealed multifocal cerebellar infarcts involving the right and left cerebellar hemispheres with no haemorrhagic conversion. A work up with ECG and telemetry demonstrated slow atrial fibrillation (AF) and a subsequent TTE highlighted a reduced ejection fraction (EF) of 25% with global hypokinesis, a dilated left ventricle (LV) and LV hypertrophy (LVH). He was initiated on appropriate medication and discharged. Ten days after his initial presentation, he re-presented with worsening dyspnoea and orthopnoea with a N-terminal pro-brain natriuretic peptide (NTproBNP) of 9605. He was treated with an acute decompensated heart failure with intravenous diuresis and medication optimisation with follow-up with the heart efficiency service. A coronary angiogram demonstrated non-obstructive coronaries.

A repeat TTE 3 months later illustrated focal severe septal hypertrophy with septal thickness up to 2.9 cm. Myocardial texture was abnormal especially in the septum and appeared bright and 'speckled'. LV systolic function was globally severely impaired with biplane left ventricular ejection fraction (LVEF) 20%, suspicious of an infiltrative cardiomyopathy (figure 1).

He attended the cardiology unit for an elective direct current cardioversion (DCCV) for AF 6 months later (10 months from index presentation). ECG at the time exhibited AF with low voltage QRS waves.

Following DCCV, the patient reverted into a sinus bradycardia with a first-degree heart block. His PR interval was significantly prolonged, with a PR interval of 300 ms prompting admission. His bisoprolol was held to prevent any bradycardic episodes. Day one post-DCCV, the patient suffered from



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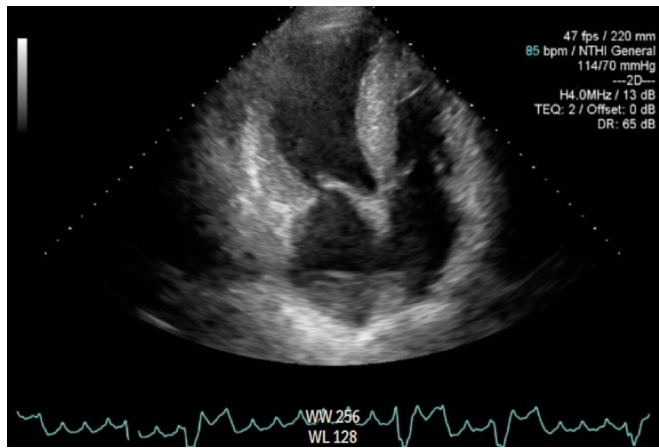


Figure 1 The patient's repeat transthoracic echocardiogram 3 months later displaying abnormal myocardial texture. The tissue appears bright and echodense suggestive of an infiltrative cardiomyopathy.

sudden onset dysarthria and confusion and urgently reviewed by the oncall medical doctor's emergency response team. On examination, he was confused and dysarthric. There was left-sided hemianopia and nystagmus present, along with dysdiadochokinesia affecting the left side more than the right. He was treated as a FAST positive stroke and he urgently underwent CT of the brain and CTA.

No acute intracranial abnormality was demonstrated with an Alberta stroke programme early CT score (ASPECTS) score of 10. There was no vascular occlusion, thrombosis or significant stenosis of the anterior or posterior circulation. Recanalisation of the previously demonstrated basilar artery occlusion was noted, along with chronic bilateral cerebellar infarcts. The patient made a full recovery with no residual neurological deficit. He was continued on apixaban and underwent MRI of the brain. Grey-white matter differentiation appeared normal. No masses or acute haemorrhage was seen. Stable periventricular deep white matter ischaemia was observable. There was evidence of some old haemorrhage within the right cerebellar hemisphere, at site of the prior infarct, which was more prominent when compared with prior. Vascular flow voids appeared patent. There was evidence of an acute infarct involving the right occipital lobe. The infarct was patchy in nature with three separate areas of infarction. Largest area of infarction measured 1.6 cm in size. He was urgently reviewed by the stroke team and diagnosed with an acute right occipital lobe infarct. During the admission, the patient recalled that he may have missed one dose of his anticoagulation 48 hours prior to his elective admission.

INVESTIGATIONS

Repeat TTE exhibited a severely dilated LV with an LV end diastolic dimension (LVEDD) of 7.1 cm. There was focal severe septal hypertrophy with septal thickness up to 2.9 cm. Myocardial texture was abnormal especially in the septum and appeared bright and echodense. LV systolic function was severely globally impaired with biplane LVEF 20%. There was no significant improvement compared with previous studies. There was mild right ventricular hypertrophy and reduced right ventricular systolic function with a dilated left atrium. The valves were grossly normal with mild tricuspid regurgitation. Right ventricular systolic pressure was elevated at 30–40 mm Hg. The TTE concluded dilated cardiomyopathy (DCM) with severe

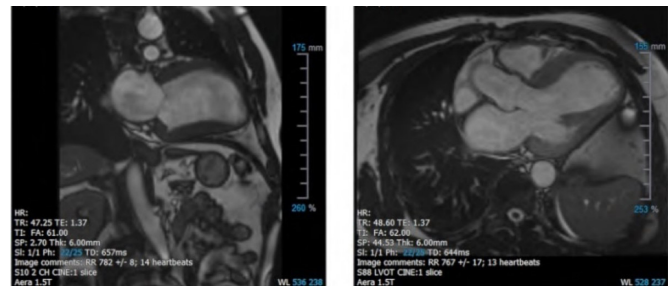


Figure 2 The patient's cardiac MRI highlighting diffuse myocardial delayed enhancement with altered gadolinium kinetics consistent with cardiac amyloidosis.

LV systolic impairment and marked focal asymmetrical septal hypertrophy.

Cardiac strain TTE was undertaken to inspect if the classic bull's eye pattern of amyloidosis was displayed. This was not visualised due to LV dilation. On contrast-enhanced TTE, no intra-cardiac thrombus was demonstrated but swirling of blood was noted in the LV. LV systolic strain was profoundly impaired with a globally reduced strain value of -2.86% .

CMR highlighted severe biventricular cardiomyopathy. The left ventricular function was estimated at 14% with swirling of the blood pool in the LV with no definite LV apical thrombus detected. The LVEDD was increased at 7.1 cm and end diastolic volume was 251 mL; global dyskinesia was shown. The right ventricle was less impaired with an EF of 32%. The left septum was again demonstrated to be asymmetrically thickened compared with the lateral wall, anteroseptal segmental thickness measured at 18 mm while the inferolateral segmental thickness was only 6 mm. There was diffuse myocardial delayed enhancement and a suggestion of altered gadolinium kinetics consistent with diffuse myocardial infiltration (figure 2). Findings were non-specific but reported as potentially consistent with CA.

Serum protein electrophoresis was normal, free kappa light chains were elevated; however, the kappa/lambda ratio was normal. Kidney function was normal with no evidence of proteinuria. Fabry's screen was negative. The patient underwent a fat pad biopsy that was positive for amyloidosis. The histology was sent to the UK for further amyloidosis subtyping. Bone marrow biopsy exhibited no infiltration.

DIFFERENTIAL DIAGNOSIS

Our patient had initially presented with sudden onset right-sided weakness, aphasia and gaze palsy with diplopia. Subsequently, he complained of worsening dyspnoea and orthopnoea with raised NTproBNP, an ECG demonstrating AF and a TTE showing reduced LV function. Our initial differential was ischaemic stroke secondary to AF and de novo non-ischaemic DCM. His subsequent TTE results raised the possibility of an infiltrative process such as amyloidosis. Other possibilities considered were medication non-compliance, Fabry's disease and multiple myeloma.

TREATMENT

Once stable, the patient's heart efficiency medication were optimised. His bisoprolol was stopped and he was commenced on sacubitril/valsartan. He was continued on apixaban.

OUTCOME AND FOLLOW-UP

Our patient was diagnosed with CA. This was based on both TTE and CMR results and confirmed with fat pad biopsy.

He made a good recovery and was discharged home, and is continuing with his heart failure therapy. Due to the ventricular dyssynchrony on TTE, a CRT defibrillator was implanted.

He is currently awaiting a serum amyloid P component scintigraphy scan at the NHS National Amyloidosis Centre in London.

DISCUSSION

Amyloidosis consists of a group of heterogeneous disorders that result from the extracellular deposition of misfolded low molecular weight proteins in abnormal fibrillar form distorting organ tissues.¹² Systemic amyloidoses can affect the renal, nervous, gastrointestinal, hematological and cardiovascular systems.^{6 13}

Cardiac involvement is the critical factor that dictates prognosis.¹⁴ Amyloid deposits in the heart produce increased ventricular wall thickness and stiffness, leading to the pathognomonic restrictive cardiomyopathy. AF's prevalence is increased in patients with CA as the amyloid protein infiltrates the atria. Atrial infiltration causes structural dissociation and increases the likelihood of atrial thrombus formation and thromboembolism, even in normal sinus rhythm.^{10 15} Deposition of amyloid in and around the arteries can lead to acute coronary syndrome.¹⁶ The free light chains can also be directly toxic to the heart by promoting cell death.¹⁷ Electrophysiological dysfunction may arise from the misfolded protein deposits, with low voltage ECG, dysrhythmias and heart block.^{18 19} During our study's timeline, our patient had both low voltage QRS complexes, AF and first-degree atrioventricular block.

Prognosis of CA is generally poor and varies depending on the specific type of amyloidosis involved.^{12 20} Based on the Mayo biomarker stage, that utilises serum cardiac troponin T, NTproBNP and serum free light chains levels, AL amyloidosis patient's median survival rates ranges from 94.1 months for stage I to 5.8 months for stage IV. AL CA patients undergoing stem cell transplantation can have median overall survival surpassing 10 years if treatment is efficacious.²¹ In contrast, transthyretin amyloidosis (ATTR) is slowly progressive and has an improved prognosis compared with AL CA.^{14 22} ATTR amyloidosis may be present in the heart of the elderly regardless of what the light chains demonstrate on protein electrophoresis.²³ The causative protein must be identified for appropriate therapy as treatments differ. Heart failure management and treatment of conduction disorders is common therapy. The mainstay of AL amyloidosis algorithms include conventional chemotherapy with agents such as cyclophosphamide, bortezomib and dexamethasone; monoclonal antibodies such as daratumumab and novel agents such as venetoclax, a BCL-2 inhibitor if the patients are found to have the translocation t(11;14).^{23 24} Based on minimal residual disease post-treatment, AL amyloid patients may be eligible for autologous stem cell transplant.^{23 24} For ATTR amyloidosis, tafamidis, a TTR binding agent, has shown a 30% reduction in all-cause mortality compared with placebo and a slower decline in 6 min walk test and in quality of life.²⁵ Other treatments include diuretics and RNA-targeted therapies such as patisiran. Heart transplantation in patients with ATTR is rare.²⁵

In the literature, ischaemic strokes in association with CA have been occasionally noted but not described well. A retrospective study in 2007 reviewed the pathophysiologic relationship between primary systemic amyloidosis and ischaemic cerebrovascular insults. The study included 40 patients with biopsy proven amyloidosis and ischaemic stroke. Thirteen of the patients' (32.5%) primary presentation was of ischaemic stroke prior to being diagnosed with systemic amyloidosis. Strokes presented 9.6 months prior to diagnosis with primary amyloidosis. Seventy per cent of patients in the study had cardioembolic infarctions.

Stroke as an initial presentation was a marker for worse prognosis, with median survival of 6.9 months following confirmation of systemic amyloidosis on biopsy. Interestingly, 37% of patients had recurrent ischaemic strokes.²⁶ In this study, this was our patient's second ischaemic stroke.

Diagnosis of CA continues to be difficult. Increased wall thickness and worsening diastolic dysfunction are two crucial characteristics on TTE of CA patients.¹ The protein deposition that causes increased LV thickness and subsequent reduced compliance leads to the diastolic failure. These features are not specific to CA and are seen in hypertrophic cardiomyopathy (HCM) and hypertensive heart disease.²⁷ However, strain imaging displays two vital differences. First, a much greater reduction in global longitudinal strain compared with HCM and hypertensive heart disease and second, a unique regional LV longitudinal strain pattern, highlighting vastly reduced basal and mid-systolic strain with apical sparing, which was shown with our patient's globally reduced strain value.²⁸ The most characteristic structural findings on echocardiogram are septal and posterior wall thickening at least >12mm, and with mean thickness of 16 mm at the time of diagnosis congruent with our patient's TTE, which demonstrated septal thickening of 29 mm.^{19 29} Classically, CA is a restrictive cardiomyopathy; however, ATTR amyloidosis can present with a reduced EF and dilation as shown with our patient.³⁰

CA increases the extracellular volume of the myocardium and in turn leads to the accumulation of gadolinium contrast.³¹ Delayed gadolinium enhancement on CMR imaging has shown to be effective in distinguishing CA. Varying non-infarct delayed enhancement patterns, from subendocardial to transmural, have been illustrated.³² CMR is extremely beneficial as an investigation to screen for CA with high specificity and positive predictive values of 80% and 81%, respectively, for diagnosis of AL CA.³³ The gadolinium contrast also displays myocardial and blood pool diskinesis deriving a reduced difference between blood and myocardium T1, a key CMR criteria.³⁴ Furthermore, the extent of the delayed enhancement by CMR is in its own a useful prognostic tool as it has been shown to be an independent predictor of mortality in AL CA.³⁵

A high frequency of intra-cardiac thrombosis are present in CA. A study at the Mayo clinic examining autopsy results of 116 CA (55 of AL and 61 of other subtypes) cases discovered intra-cardiac thrombus in 33%. Twenty-three had 1 thrombus, whereas 15 had multiple; for a total of 63 thrombi. The AL subgroup had notably increased intracardiac thrombus found at autopsy (51% vs 16%) and additional fatal embolic events (26% vs 8%) when compared with the other amyloidosis subgroups.¹⁰ The authors identified that if AL amyloidosis subtype and AF were simultaneously present, there was a substantially higher risk for thromboembolism.¹⁰ In this study, although our patient showed no evidence of intra-cardiac thrombus on TTE or CMR, he did have both CA and AF increasing his risk for thromboembolism as evident by the recurrent cerebral ischaemic events. Though TTE is a key non-invasive tool for cardiac imaging, transesophageal echocardiogram is the superior modality for the investigation of many cardiac sources of emboli,^{36 37} with CMR being more sensitive for left ventricular thrombus detection.³⁸

The mechanisms of thromboembolism in CA are multifactorial. Mural thrombosis may develop due to blood stasis and intracavitary turbulence as seen in our patient's strain TTE and CMR.³⁹ Deposition of amyloid to endocardial tissue has shown to cause fibrous thickening and impaired wall kinetics contributing to endocardial thrombi.⁴⁰ Dysrhythmias as stated previously is another well-documented cause of cerebral ischaemia. The prevalence of AF in CA is approximately 20%. It is theorised

Rare disease

that this is due to left atrial failure secondary to dilation and raised wall stress.⁴⁰ Using 24 hours hotter monitoring, the incidence of ventricular arrhythmias in amyloidosis was recently evaluated. The study highlighted a significant incidence of arrhythmias, including ventricular ectopies in 72% and complex ventricular arrhythmias in 57% of patients.⁴¹ Amyloidosis also creates a hyper-coagulable state either from damage of the renal system through proteinuria, infiltration of the spleen causing thrombocytosis, the direct effect on the coagulation cascade or increased blood viscosity or procoagulant activity associated to the circulating monoclonal component all also increase the risk for thromboembolism.⁴²

Patient with amyloidosis can also present with carpal tunnel syndrome which may be bilateral. A study of 98 patients undergoing carpal tunnel release surgery, underwent simultaneous Congo red staining of tenosynovial tissue. This subsequently detected amyloid deposits in 10 of the patients (7 ATTR, 2 AL, 1 untyped). Concomitant cardiac evaluation identified two patients with involvement of the myocardium (1 ATTR, 1 AL), allowing for implementation of disease-modifying therapy.⁴³ This has led to the possibility of screening patients with carpal tunnel syndrome amyloidosis. Interestingly, our patient also suffered from carpal tunnel syndrome.

Learning points

- ▶ Stroke is a potential complication of systemic amyloidosis, and can also be the initial presentation. Stroke as an initial presentation was a marker for worse prognosis; therefore, early diagnosis is imperative.
- ▶ The diagnosis of cardiac amyloidosis (CA) should be considered in patients with cardiac dysfunction, left ventricular hypertrophy and a speckled appearance of myocardium on transthoracic echocardiogram (TTE).
- ▶ There is an intrinsic tendency towards thromboembolism with amyloidosis. CA patients may have recurrent ischaemic strokes, especially if AF is simultaneously present. It is imperative that strict adherence and compliance to anticoagulation regimes are met, as this may reduce morbidity and mortality.
- ▶ Investigations in these groups of patients suspicious for CA include TTE, cardiac MRI, subcutaneous fat biopsies and if warranted endomyocardial biopsy to aid in early diagnosis and treatment of CA.

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