Cardiac involvement in systemic inflammatory diseases

Daniel C. Knockaert

Department of General Internal Medicine, University Hospital Gasthuisberg, Herestraat 49, 3000 Leuven, Belgium

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Introduction

Systemic inflammatory diseases (SIDs) can be defined as inflammatory syndromes with constitutional symptoms and involvement of at least two organ systems. Systemic lupus erythematosus (SLE) is the most impressive and typical example of an SID. However, not all patients presenting with systemic symptoms have an underlying SID. Knowledge of the broad array of conditions that may mimic SID with cardiac involvement is essential because specific and causal treatment can be given for a number of them (Table 1). Bacterial endocarditis, acute rheumatic fever, and Lyme disease, represent the best known examples of infections suggesting an SID. Most cardiologists are not so familiar with SIDs because major cardiac problems are rarely presenting manifestation. Cardiac involvement presents either as: (i) pericarditis; (ii) myocarditis or myocardial fibrosis due to myositis or vasculitis with rhythm and conduction disturbances and diastolic or systolic heart failure; (iii) coronaritis with ischaemic heart disease; (iv) endocardial involvement with valvular disease and formation of thrombi; (v) pulmonary hypertension secondary to concomitant lung disease or recurrent lung embolism; (vi) unexplained arterial thrombosis; (vii) syncope; and (viii) (malignant) arterial hypertension.1 Cardiomyopathy is common in several vasculitides and it is caused by either diffuse myocardial ischaemia due to vasculitis of small epicardial vessels or by granulomatous or eosinophilic infiltration depending upon the type of vasculitis. I also included thoracic aortic aneurysm and dissection as cardiac involvement because most of these patients are treated by cardiologists and cardiac surgeons. The reported frequency of cardiac involvement in SIDs ranges widely depending upon the applied diagnostic methods and selection of patients. Autopsy studies revealed a much higher frequency of cardiac involvement than clinical case series in the older literature. The introduction of transoesophageal echocardiography, sophisticated myocardial perfusional scintigraphy techniques, and magnetic resonance imaging (MRI) has resulted in prevalence figures close to and even higher than those reported in the older autopsy studies.2 The frequency of cardiac findings is rather low at the time of diagnosis in many SIDs, yet considerably higher during follow-up.3,4

Search strategy

MEDLINE database was searched for the period January 1, 1990 through September 2005, using PubMed for articles containing ‘cardiac’, ‘cardiovascular’, ‘heart disease’, and the SIDs discussed in this review. We did not include data from older studies because many definitions and diagnostic criteria have changed since the early 1990s. The search was limited to English language studies.

Large case series were used to determine the frequency of cardiac manifestations and case reports were included in the reference list only when they described unusual features.

Takayasu’s arteritis

This is more common in Asians than in other racial groups, the female-to-male ratio is 10:1, and more than 90% of the patients are <40 years of age. These epidemiological features represent the cornerstone for the differential diagnosis with temporal arteritis. The typical distribution of vessel involvement is the thoracic aorta itself but also the abdominal aorta and the proximal parts of its major branches. It is a panarteritis with inflammatory wall thickening evolving towards a fibrotic obliterative stage with aneurysms, stenosis, and occlusion.5
Involvement of the coronary arteries occurs in 15–25% of cases, most commonly near the ostium and less frequently in the distal portions of the coronary arteries. Aneurysm of the coronary arteries and fistulas between coronary artery and bronchial artery are rare manifestations. Aortic regurgitation develops as a consequence of aneurysmal dilatation of the proximal aorta. Pulmonary arteries are affected in 15–70% of all patients and pulmonary artery stenosis was found in four of 60 patients. This causes pulmonary hypertension and right heart failure. Pulmonary artery aneurysm may present as hilar enlargement and cause life-threatening lung bleeding. Fistulas can develop between pulmonary artery and bronchial arteries, coronary arteries, or aorta. Subclinical myocardial involvement was found in 10 of 21 patients without significant coronary artery lesions. Pericardial effusion is uncommon, but may even be the initial manifestation. Renovascular involvement occurs in more than half of the patients and malignant hypertension may be the initial presentation.

**Giant cell arteritis**

Giant cell arteritis (GCA), also called temporal or granulomatous arteritis is the most common vasculitis in populations with Northern European ancestry. This large vessel vasculitis seldom presents below the age of 50. It rarely causes cardiac problems but the considerably increased relative risk, up to 17 times, to develop thoracic aortic aneurysm and aortic dissection is poorly known. This was initially reported in a population-based cohort study of 96 patients of whom 11 had thoracic aortic aneurysm. The aneurysm was detected at the time of diagnosis in two patients and in the remaining nine cases after a median follow-up of 5.8 years. These findings were confirmed by the same group in a larger study of 168 patients and by other investigators who reported thoracic aortic aneurysm and/or dissection in 16 (7.6%) of 210 patients. GCA can cause coronary artery and myocardial infarction but it is not known how often this occurs. Pericarditis has been described in several case reports.

**Polyarteritis nodosa**

Polyarteritis nodosa (PAN) was historically divided on a clinical basis as classic PAN and microscopic PAN. The latter has been renamed as microscopic polyangiitis (MPA) and separated from PAN according to the size of involved vessels. Classic PAN is a necrotizing arteritis of medium-sized and small arteries without involvement of arterioles, capillaries, or venules. In a study on prognostic factors in PAN (n = 260) and Churg–Strauss syndrome (n = 82), cardiomyopathy was the sole type of cardiac involvement mentioned, present in 15 of the 342 cases. Coronary vasculitis causing multiple aneurysms and myocardial infarction has been described in several case reports of PAN. Severe or malignant arterial hypertension is a classic manifestation of PAN, caused by renal artery vasculitis.

**Microscopic polyangiitis**

Microscopic polyangiitis (MPA) always affects small vessels (capillaries, venules, arterioles) but medium-sized vessels may be involved. In a series of 85 patients a surprising high frequency (50.6%) of cardiovascular manifestations was found. Pericarditis, heart failure, and hypertension was present in respectively 9, 15, and 29 cases and myocardial infarction in two. In a retrospective series of 72 patients with PAN (n = 36) and MPA (n = 36), 12 individuals (17%), six in each group, had evidence of cardiac involvement at the time of diagnosis. Takatsubo (ampulla-shaped apical ballooning and hypokinesis) cardiomyopathy has been described in a 70-year-old patient with active MAP and complete recovery after treatment with corticosteroids.

**Kawasaki disease**

Kawasaki disease (KD) is a vasculitis of medium-sized vessels and probably the vasculitis best known by cardiologists. The majority of cases occurs in children under 5 years of age. Rare cases have been reported in adult patients, but most cases probably are due to late sequelae and not due to recent onset active disease. The incidence is much higher in Japan than in Western countries. The dreaded cardiac complication is coronary artery involvement, present in 20% of cases, resulting in myocardial infarction and aneurysm formation. Myocarditis is probably universal in the acute stage and this causes arrhythmia, heart failure, and valvular abnormalities. Pericardial effusion is found in 30% of cases. The sequelae of KD can result in myocardial infarction and death due to coronary artery occlusion several years after the onset of the disease. Hence, KD should be considered as the possible cause of acute coronary syndromes in young adults, particularly when aneurysms are found.

### Table 1 Differential diagnosis of diseases with systemic presentation and cardiac involvement

<table>
<thead>
<tr>
<th>Systemic inflammatory diseases (SIDs)</th>
<th>Vasculitis, connective tissue diseases, granulomatous diseases</th>
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</thead>
<tbody>
<tr>
<td>Infections</td>
<td>Bacterial, tuberculosis, Rickettsia and Bartonella, Erlichia, Anaplasma, parasites, viruses</td>
</tr>
<tr>
<td>Toxins</td>
<td>Substance abuse (cocaïne, amphetamines, ecstasy), lead poisoning, paraquat poisoning</td>
</tr>
<tr>
<td>Medications</td>
<td>Drug-induced hypersensitivity syndrome, drug-induced lupus erythematosus, chloroquine, ergotamine, bromocrypentine, pergolide, amiodarone, anorectic drugs</td>
</tr>
<tr>
<td>Infiltrative or storage disease</td>
<td>Haemochromatosis, amylodosis, lysosomal storage disorders (Gaucher’s disease, Fabry’s disease), glycogen storage diseases, hyper-eosinophilic syndrome, mastocytosis, Erdheim–Chester disease, Refsum’s disease</td>
</tr>
<tr>
<td>Hereditary diseases</td>
<td>Familial Mediterranean fever, Marfan syndrome, Ehlers–Danlos syndrome, Pseudoxanthoma elasticum, primary mitochondrial diseases</td>
</tr>
<tr>
<td>Other</td>
<td>Atrial myxoma, cholesterol embolism, cryoglobulinaemia, primary antiphospholipid syndrome (Hughes syndrome), inflammatory bowel disease, multifocal fibrosclerosis Rosai-Dorfman disease</td>
</tr>
</tbody>
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absence of generalized atherosclerotic disease, a history of KD-like illness in childhood should always be sought in these cases. Follow-up of children with KD should be individualized according to the risk of late myocardial ischaemia. This risk is determined by the presence, type of coronary artery abnormality, and the change of these abnormalities over time.23

Wegener disease

Wegener disease (WD) is a necrotizing vasculitis of medium and small size vessels with granulomatous lesions classically involving the upper and lower respiratory tract and the kidneys.25 Pericarditis, coronary arteritis, aortic regurgitation, and aortic valvular lesions simulating endocarditis are the most commonly reported abnormalities.26 Heart block and aortic aneurysm have been described in case reports. Pericarditis has been documented in 6% of patients with WD in older case series.25 Cardiac manifestations were present at baseline in 20 of 155 patients diagnosed in the period 1966–93.27 A retrospective study of 85 consecutive patients with WD, referred to the echocardiography laboratory, reported echocardiographic abnormalities in 73 (86%) individuals but lesions were considered as related to WD only in 26 (30%).28 Pericardial effusion, regional wall motion abnormalities, and left ventricular (LV) systolic dysfunction were found in respectively in 5, 17, and 13 cases. This high number probably reflects referral bias and clinical cardiac manifestations were rather rare in large cohorts of patients with WD.25,27,29 Valvular disease, mainly aortic valve insufficiency, is probably more common than generally accepted.26

A recent study revealed an increased risk of thrombo-embolic events in patients with WD but pulmonary hypertension is exceptionally described up till now.30

Churg–Strauss syndrome

Churg–Strauss syndrome is characterized by necrotizing small-sized vessel vasculitis, extravascular granulomas, and eosinophilia. The typical clinical manifestations are pulmonary infiltrates and mononeuritis multiplex in patients with rather severe asthma.31 Cardiac involvement is common and a leading cause of mortality with a frequency ranging from 16 to 92%.32 Myocarditis and coronary arteritis account for approximately 50% of deaths if the disease remains unrecognized.33

Coronary involvement has rarely been found pre-mortem in contrast to necropsy studies.33 Eosinophilic endomyocarditis and fibrosis with thrombus formation, similar to what is observed in idiopathic hypereosinophilic syndrome, is extremely rare.34 Pericardial effusion was detected at the time of diagnosis in 21 of 96 patients (22%), heart failure in nine, and transient heart block in three cases.35 In another study, 35% of 112 patients had cardiac manifestations. Pericarditis was present in 28 cases (25%) with tamponade in seven cases, cardiomyopathy in 27 cases (24%) of whom 20 (18%) had heart failure. Heart disease was inversely associated with ANCA positivity; 49% of ANCA negative patients had cardiac manifestations in contrast to 12% of ANCA positive cases.35

Systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is probably the best known SID.16,37 All components of the heart can be affected and venous thrombo-embolic complications may cause pulmonary hypertension and right heart failure.38

Symptomatic pericarditis occurs in about 25% of the patients and asymptomatic pericardial involvement in more than half of the cases.38 Older autopsy studies reported pericardial involvement in up to 80% of the patients. Tamponade occurs in less than 2% and constrictive pericarditis is extremely rare. Cardiomyopathy is uncommon clinically but autopsy studies found myocardial involvement in 40–50% of patients. The relationship to SLE is not always clear because hypertension, atherosclerosis, and even anti-malarials such as chloroquine used in the treatment may play a role.39,40 Myocarditis in contrast to pericarditis is an indication for treatment with corticosteroids.

Valvular disease has initially been described in 1924 as Libman-Sacks or marantic endocarditis. It consists of non-infective, verrucous vegetations and it represents the most characteristic valve lesion in SLE. Any valve may be involved but most frequently the mitral and to a lesser degree, the aortic valve and multivalvular involvement is common. Selection of patients and echocardiographic technique partly explains the great variance in reported incidence of valvular disease.

The landmark study of Roldan et al.41 described valvular abnormalities in 61% of 69 cases. Valvular disease was not related to disease severity, disease activity, or disease duration. An older prospective study of 132 consecutive patients reported valvular lesions in 22.7% of the cases and valvular lesions were associated with the presence of anti-phospholipid antibodies.42

Neonatal lupus syndrome is an in utero acquired autoimmune disease, characterized by congenital heart block. It occurs in a small proportion of children born to mothers with anti-RO/SSA and anti-LA auto-antibodies.36

Coronary involvement is rare but coronary dissection and coronary artery aneurysm may occur.43,44

Accelerated or premature atherosclerotic disease is a leading cause of late death in patients with SLE, while active disease and severe infections are more important causes of death in the early phase of the disease.37,45–48 The pathogenesis and risk factors of this premature atherosclerosis in a disease that mainly affects young women, a group usually free of atherosclerosis, are not fully understood. Coronary vasculitis is not considered to be the underlying mechanism but persistent inflammation, autoimmunity, immune complex deposition, and antiphospholipid antibodies are hypothesized to cause intimal damage followed by accelerated atherosclerosis.37,47 A role of duration of treatment with corticosteroids has initially been suggested but not proven. Corticosteroids indeed have adverse effects on traditional risk factors for atherosclerosis such as blood pressure, lipid profile, obesity, and blood glucose level.46 Several investigators suggest rather that chronic inflammation is atherogenic and the association of glucocorticoid use with premature atheromatosis might be a reflection of severity of the inflammation requiring higher doses of glucocorticoids.45,48

Similar findings have been reported in patients with rheumatoid arthritis49–51 and in patients with temporal arteritis.52
Systemic sclerosis

Systemic sclerosis (SSc) scleroderma, is characterized by excessive production of collagen resulting in relentless fibrosis of the involved organs. Female-to-male ratio is 7–10:1.53,54 Cardiac involvement is one of the factors shortening survival but patients with renal involvement have the worst 10-year survival rate.54,55 The kidneys are particularly at risk in the so called scleroderma renal crisis, characterized by severe hypertension, rapidly increasing serum creatinine concentration, and microangiopathic haemolytic anaemia. Prompt aggressive treatment with ACE-inhibitors can reverse this otherwise fatal process.56

Cardiac involvement consists mainly of pericardial and myocardial disease causing diastolic dysfunction, congestive heart failure (CHF), severe ventricular arrhythmia and atiroventricular conduction abnormalities. Thickening of valve leaflets is less frequent but right heart failure is common and mostly secondary to lung disease. An autopsy study of 44 patients revealed myocardial fibrosis in 37% of the cases and chronic pericarditis in 72%.57 Cardiac involvement, defined as pericarditis, heart failure, arrhythmia and/or conduction abnormalities, was found in 30% of the cases in a large study.54 An echocardiographic study of 77 consecutive patients revealed pericardial abnormalities in 43% of the cases, LV diastolic dysfunction in 29%, tricuspid regurgitation in 40%, thickening of the aortic valve and mitral valve leaflets in, respectively, 12 and 8%, and pulmonary hypertension in 47%.58 The latter is mostly secondary to lung involvement and these patients have a 1 year survival rate of only 50%. Several cases of massive pericardial effusion and tamponade have been reported.

Coronary involvement is not a feature of SSc but coronary flow reserve was significantly lower in a group of 27 patients with SSc than in controls.59

Limited cutaneous SSc, previously called CREST (calcinosis, Raynaud, oesophageal dysmotility, sclerodactyly, telangectasia), has a more favourable outcome. However, it causes pulmonary hypertension, mostly without concomitant interstitial lung disease in 10–15% of the cases. Cardiac symptoms are rare in limited SSc but subclinical LV dysfunction as evidenced by echocardiography and perfusion scintigraphy was found in 42% of 19 patients and mild pericardial effusion in two cases (10%).60

Polymyositis and dermatomyositis

Polymyositis (PM) and dermatomyositis (DM) are rare idiopathic inflammatory myopathies.61 Inclusion body myositis, the third and most common inflammatory myopathy in patients older than 50 years, seems not to affect the heart.62 Tachyarrhythmia, conduction disturbances, myocarditis, and CHF are the classical cardiac manifestations and, when present, associated with a poor prognosis. Older autopsy studies revealed cardiac abnormalities in up to 40% of the patients.63 A detailed cardiologic assessment of 32 patients with DM/PM revealed cardiac symptoms in only two cases but conduction disturbances in more than half of the patients and LV diastolic dysfunction in 42% of the cases studied by echocardiography.63 ECG abnormalities were described in 17.3% of 75 patients with PM/DM.64 Cardiac involvement was reported in 9.3% of 162 cases with idiopathic inflammatory myopathy followed-up for a minimum of 5 years but it might have been under-diagnosed due to the retrospective design of the study.65 Cardiac tamponade has been described in case reports.

Interstitial lung disease is one of the most common extramuscular manifestations found in up to 5–30% of the cases. This may lead to right heart failure.66,67

Mixed connective tissue disease

Mixed connective tissue disease is a so-called overlap disease with features of systemic sclerosis, SLE, PM/DM, and rheumatoid arthritis.68,69 Cardiac findings are comparable with the cardiac manifestations, described in the diseases that constitute the overlap syndrome. Pericarditis was found in 29% of the cases in older studies and in 56% of autopsied patients.68,69

Pulmonary hypertension is the most serious complication and leading cause of death.68 It was found in 23% of 47 patients followed-up at a specialized referral centre.70

Primary Sjögren syndrome

Primary Sjögren syndrome (SS), keratoconjunctivitis sicca, is an autoimmune disease with lymphocytic infiltration of the salivary and lacrimal glands. When associated with other SIDs it is called secondary SS.71–73 It occurs mostly in the fourth to sixth decade and the female-to-male ratio is 9–14:1.

Clinically obvious cardiac disease is uncommon and was not mentioned in a recent series of 400 patients.73 However, another study reported silent echocardiographic evidence of pericardial involvement in 21 (33%) of 64 patients with primary SS,74 a figure similar to that of another small study of 27 patients.75 Evidence of diastolic dysfunction was found in 21 of 42 evaluated patients and this was independent of the pericardial findings.74 Myocarditis is extremely rare.76 Congenital heart block, associated with the presence of anti-SS A Ro-antibodies, a problem well known in patients with SLE, also occurs in primary SS.77

Lung involvement may cause secondary pulmonary hypertension and pulmonary pressure was significantly greater in patients with primary SS than in the controls.74

Adult-onset still disease

Adult-onset still disease (AOSD) typically causes episodic high fever, transient and difficult to see erythema, throat pain, joint symptoms, lymphadenopathy, and splenomegaly. Pericarditis is present in 20–37% of the cases78,79 and myocarditis was found in 10%,79

Relapsing polychondritis

Relapsing polychondritis is an extremely rare multisystem disease with recurrent inflammation and destruction of cartilaginous tissues throughout the body, particularly external auricular, nasal, and laryngo/tracheal cartilage, joints, and other structures.4

Cardiac pathology is rather rare at presentation but it develops at a mean of 6 years after the onset of clinical symptoms in 10–22% of the cases.80–82 Cardiac involvement is the second most frequent cause of death, pneumonia being the most common.4,82 Older studies report
cardiovascular involvement in up to 56% of cases but vasculitis and thrombosis was included in that figure. In more recent studies frequency of cardiac manifestations varies between 10 and 25%.82

The classic cardiac complication is valvular disease, particularly aortic regurgitation which is more common (4-10% of cases) than mitral regurgitation (2-4%).4,80 A multicenter study of 62 unselected patients reported cardiac abnormalities in 22% of the patients but not all abnormalities were considered manifestations of relapsing polychondritis.82 Four patients had pericarditis, two had atrioventricular block, five had re-entry tachycardia, three had coronary heart disease but, surprisingly, valvular disease was not mentioned. Aneurysms develop in 5-7% of patients along the entire length of the aorta, more often in the ascending aorta and also in the coronary arteries.30,83

Behçet’s disease

Behçet’s disease (BD) is a relapsing inflammatory disease with recurrent aphthous stomatitis, genital ulcerations, and uveitis as most typical manifestations. Its prevalence is high along the ancient Silk trade Route (from the eastern Mediterranean littoral to Japan) but very low in the US and Western countries.84

Cardiac involvement is found in only 1-5% of patients in older series but vascular abnormalities, both arterial and venous, develop in up to 50% of patients.85 Myocarditis, atrioventricular block, pericarditis, coronary and valvular involvement, particularly of the aortic valve, and intracardiac thrombosis (right atrial and ventricular thrombosis), represent the cardiac pathology.84 Aortitis and aortic regurgitation are considered to be rare86 but BD was found to be the cause of aortic regurgitation in seven of 153 consecutive patients requiring aortic surgery in a Korean series.7 Valve surgery is frequently complicated by suture or valve detachment.87 Coronary artery involvement occurs in <1% of cases and coronary artery bypass grafting is difficult because of tissue fragility and there is also a risk of pseudoaneurysm formation. Hence, a rather conservative approach is proposed in case of symptomatic coronary artery disease.88 Vascular abnormalities, both arterial and venous are much more frequent than cardiac involvement.84 Aneurysms, particularly of the pulmonary arteries, known as the Hughes–Stovin syndrome and also of the coronary and peripheral arteries are a typical feature of BD. It is well known that arterial puncture may provoke the development of aneurysms at the puncture site. Venous thrombosis are so common that BD is included in the differential diagnosis of thrombophilia. Endomyocardial fibrosis is rare and it predominantly involves the right ventricle.89

Cogan’s syndrome

Cogan’s syndrome is one of the rarest and least known systemic diseases with aortic aneurysm and aortic insufficiency as serious cardiac manifestation. It is mainly a disease of children and young adults with median age at onset of 25 years. The typical features are interstitial keratitis and vestibulo-auditory dysfunction. Systemic symptoms develop in half of the cases.90,91 It affects typically the thoracic aorta but also medium- and small-sized arteries. Other reported cardiovascular manifestations in Cogan’s syndrome are CHF, pericarditis, coronary vasculitis, and atrioventricular block.92

The differential diagnosis of inflammatory disease of the aorta includes temporal arteritis, Takayasu’s arteritis, ankylosing spondylitis, Behçet disease, Cogan’s disease, and syphilis although SLE, rheumatoid arthritis, sarcoidosis, and others may occasionally be the cause (Table 2).

Sarcoidosis

Sarcoidosis can be considered as an SID, histologically characterized by non-caseating granulomatous inflammation. Patients may be entirely asymptomatic and 30-60% are discovered on routine health screening chest radiograph.93,94 Ventricular and supraventricular arrhythmia, conduction disturbances, restrictive or CHF, syncope, and sudden death are the typical manifestations of myocardial involvement, summarized in recent reviews.2,95-97

Patients in whom myocardial involvement is thought to be likely should be assessed by echocardiography, Holter monitoring, myocardial scintigraphy, or 18 FDG PET. Gadolinium-enhanced MRI has recently emerged as the most sensitive and specific diagnostic tool to determine cardiac involvement.

Conclusion

The true prevalence and clinical importance of cardiac abnormalities in most SIDs, both at the time of presentation and during evolution of the disease, is difficult to delineate.

Table 2 Causes of thoracic aneurysm

<table>
<thead>
<tr>
<th>Cause</th>
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<tbody>
<tr>
<td>Atherosclerosis</td>
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<tr>
<td>Infectious vasculitis</td>
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<tr>
<td>Syphilis, Salmonella species, tuberculosis</td>
</tr>
<tr>
<td>Primary vasculitis</td>
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<tr>
<td>Takayasu arteritis</td>
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<tr>
<td>Temporal arteritis</td>
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<tr>
<td>Polyarteritis nodosa (PAN)</td>
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<tr>
<td>Wegener’s granulomatosis</td>
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<tr>
<td>Connective tissue diseases</td>
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<tr>
<td>Relapsing polychondritis</td>
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<tr>
<td>Cogan’s syndrome</td>
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<tr>
<td>Ankylosing spondylitis</td>
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<tr>
<td>Behçet’s disease</td>
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<tr>
<td>Rheumatoid arthritis</td>
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<tr>
<td>Systemic lupus erythematosus (SLE)</td>
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<tr>
<td>Hereditary diseases</td>
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<tr>
<td>Ehlers–Danlos syndrome</td>
</tr>
<tr>
<td>Marfan’s syndrome</td>
</tr>
<tr>
<td>Pseudo-xanthoma elasticum</td>
</tr>
<tr>
<td>Osteogegenes imperfecta</td>
</tr>
<tr>
<td>Miscellaneous</td>
</tr>
<tr>
<td>Sarcoidiosis</td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
</tr>
<tr>
<td>Thromboangitits obliterans (Buerger disease)</td>
</tr>
<tr>
<td>Multifocal fibroclerosis</td>
</tr>
<tr>
<td>Fibromuscular dysplasia</td>
</tr>
<tr>
<td>Drugs (ergotamine, methysergide, etc.)</td>
</tr>
<tr>
<td>Radiation</td>
</tr>
<tr>
<td>Erdheim–Chester disease</td>
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<tr>
<td>Idiopathic</td>
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from the existing literature data. Sophisticated technological assessment reveals frequencies of cardiac abnormalities similar to and even higher than the older autopsy studies for several SIDs. However, the clinical significance of these findings is not yet established. Prospective studies of unsel ected and strictly defined cases, using appropriate techniques and correlation with clinical and autopsy data, are highly needed to answer these questions. Better knowledge and awareness of cardiac involvement in SIDs is necessary because it conveys a major risk for increased morbidity and mortality in several of these rare diseases. This has clearly been shown for temporal arteritis, Churg–Strauss syndrome, sarcoidosis, SSC, relapsing polychondritis and others.

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Conflict of interest: none declared.

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12. Salvarani C, Cantini F, Boiardi L, Hunder GG. Polymyalgia rheumatica in several of these rare diseases. This has clearly been shown for temporal arteritis, Churg–Strauss syndrome, sarcoidosis, SSC, relapsing polychondritis and others.

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**Clinical vignette**

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Spontaneous intramural gastric haematoma as a complication of oral anticoagulant therapy mimicking acute myocardial infarction

Laurent Leborgne*, Amel Mathiron, and Geneviève Jarry

Cardio Vascular Center, University of Picardie, Amiens, France

* Corresponding author: Service de Réanimation Cardiaque, CHU AMIENS, 80054 Amiens Cédex 01, France. Tel: +33 3 22 45 58 70; fax: +33 3 22 45 56 61.

E-mail address: leborgne.laurent@chu-amiens.fr

A 73-year-old patient was admitted for acute pain in the low retrosternal area and nausea. He was a smoker and had a history of myocardial infarction and paroxystic atrial fibrillation treated with fludione, amiodarone, and aspirin. At time of admission, the patient had a sinus tachycardia, and blood pressure was 100/45 mmHg. Initial ECG showed Q-waves and persistent ST-elevation in opposite leads (Panel A). Diagnosis of recurrent myocardial infarction was considered and patient was transferred to the catheterization laboratory. Coronary angiography was performed, which revealed a chronic total occlusion of the left anterior descending coronary artery (Panel B) with a very good collateral flow (Rentrop grade 3) (Panel C), excluding an ischaemic cause of the current symptomatology. Patient became shocked, and meanwhile, haematology laboratory alerted us that haemoglobin was 8.6 g/dL and INR was 12.1. A gastroscopy was rapidly performed, which showed no sign of active bleeding. The abdominal multi-slice computed tomography (MSCT) revealed a spontaneous and voluminous intramural haematoma localized in the greater curvature of the stomach (Panel D), associated with a small haematic peritoneal effusion, and intraperitoneal fat infiltration. The patient recovered rapidly after blood transfusion, volumic expansion, IV vitamin K, and IV PPSS-concentrate. One year later, the abdominal CT showed only a small remnant haematoma (Panel E).

Even if it has been formerly reported that a significant number of patients treated with anticoagulant had jejunal and duodenal submucosal haemorrhages at autopsy, clinically relevant anticoagulant-induced intramural haematomata of gastrointestinal tract is very rare and only one case of gastric localization is already related in the literature. The treatment of choice is conservative with correction of coagulation disorders. Surgery should be reserved to patients exhibiting clinical signs of gastric necrosis or peritonitis. Moreover, this case underlines the importance of not using thrombolytic or aggressive antiplatelet therapies before coronary angiography (to confirm the diagnosis) in patient with acute myocardial infarction previously treated with oral anti-coagulant.

Panel A. ECG showing peaked T-waves, ST-elevation, and Q-waves in antero-septal leads (V1, V2, V3, V4).

Panels B and C. Coronary angiogram depicted proximal occlusion of the left anterior descending coronary artery (Panel B, arrow) with a Rentrop 3 collateral flow (Panel C, black arrow) coming from the right coronary artery (Panel C, white arrow).

Panels D and E. Abdominal MSCT showing large intramural haematoma in the greater curvature of the stomach (Panel D, long arrow), with small haematic peritoneal effusion (small arrow), and intraperitoneal fat infiltration (arrowhead). Small remnant haematoma at 1 year (Panel E, arrow).