Unusual association of diseases/symptoms

Myopericarditis in giant cell arteritis: case report of diagnostic dilemma and review of literature

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Giant cell arteritis (GCA), also known as granulomatous arteritis is a systemic vasculitis mainly affecting extra cranial branches of carotid arteries. It can rarely affect other vascular beds causing thoracic aorta aneurysm, dissection and rarely cause myocardial infarction through coronary arteritis. It can cause considerable diagnostic dilemma due to varied clinical presentations. The authors report an illustrative case of a 70-year-old woman with GCA who developed symptoms suggestive of acute myocardial infarction with chest pain, localised ST-T changes and echocardiographic left ventricular dysfunction. However, cardiac troponin T biomarkers and coronary angiography were normal. Her symptoms subsided with steroid treatment. Cardiac symptoms at first presentation of GCA are unusual.

BACKGROUND

Giant cell arteritis (GCA), also known as temporal arteritis is a systemic vasculitis, known to affect the extra cranial branches of carotid artery and involves extracranial arteries in approximately 10% cases.¹ It can cause segmentary vascular localisation and can cause thoracic aortic aneurysm, dissection, aortic stenosis, aortic valvular insufficiency and rarely, myocardial infarction through coronary arteritis but the incidence is unknown. Pericarditis and myopericarditis are rarer manifestations that have been described in case reports but myopericarditis as a mimic of myocardial infarction due to limited vascular territory localisation on ECG and left ventricular (LV) dysfunction has not been described.

Several conditions can present with chest pain. These conditions may at times behave like mimics of acute coronary syndrome (ACS). This can cause dilemma in instituting appropriate treatments in the emergency room. Sometimes, manifestations of GCA can behave like an ACS mimic. Here, we present a case of a 70-year-old woman with GCA who presented with symptoms and ECG changes suggestive of acute myocardial infarction (AMI), LV dysfunction and repeatedly normal cardiac biomarker (cardiac troponin T) causing considerable initial diagnostic dilemma. We will also review available literature.

CASE PRESENTATION

A 70-year-old lady presented to the accident and emergency department with a sudden onset of precordial pain. She described it as a sharp stabbing pain, radiating to the left shoulder and arm, associated with sweating and breathlessness.

She was previously investigated a month before by the surgeons at the hospital for normocytic anaemia and weight loss. Various investigations including gastroscopy, colonoscopy and CT scan of chest/abdomen/pelvis were performed with normal results and no cause for anaemia was found.

At presentation, her heart rate was 100/min with ectopic beats, blood pressure 150/70 mm Hg, temperature 36.7°C and examination of cardiovascular, chest, abdominal and nervous systems were unremarkable.

The ECG showed sinus tachycardia, biphasic and fluctuating anterior ST elevation and saddle-shaped ST elevation in the inferior and lateral leads (figure 1). As there was some uncertainty in her diagnosis, she was not given intravenous thrombolytic treatment. The cardiac troponin T was normal, but a subsequent echocardiogram showed an enlarged LV cavity, lower septum and apical marked hypokinesia, LV ejection fraction of 40%, but no significant pericardial effusion (figures 2 and 3). This was suggestive of a myocardial infarction or myopericarditis, albeit with normal cardiac enzymes.

Figure 1 (A) ECGs on admission and changes over initial 5 days. (B) ECG 1 year later.

Figure 2 Sequential echocardiography – apical four chamber view (LV, left ventricle; LA, left atrium; RA, right atrium). (A) Three days later – lower septum and apical hypokinesia (AH), left ventricular ejection fraction of 40% and no significant pericardial effusion, (B) 6 weeks later – improvement in LV function and development of small pericardial effusion (PerE), (C) 7 weeks later – large pericardial effusion predominantly over right heart (arrows pointing to PerE), (D) 3 months later – no pericardial effusion.

Figure 3 Sequential echocardiography – subcostal view (LV, left ventricle, LA, left atrium, RA, right atrium). (A) Three days later – no significant pericardial effusion, (B) 6 weeks later – small pericardial effusion (PerE), (C) 7 weeks later – large pericardial effusion especially over right heart, (D) 3 months later – no pericardial effusion.

The inflammatory markers were elevated with erythrocyte sedimentation rate (ESR) 89 mm/h and C reactive protein (CRP) 69 mg/l. She had also complained of inflammatory stiffness in the shoulder girdle in the preceding week and it was thought she might have polymyalgia rheumatica (PMR). She was therefore commenced on 15 mg of prednisolone per day. She underwent a coronary angiogram a few days later which showed normal coronary anatomy

and no evidence to suggest recent acute plaque rupture and also improvement in cardiac function with normal ejection fraction of 65% (figure 4). Her symptoms did settle with the prednisolone and she was discharged home on these steroids with a presumptive diagnosis of possible viral myopericarditis and PMR. However, viral serology for cardiotropic viruses was negative. A cardiac biopsy was not considered due to normalisation of cardiac functions noted

Figure 4 Coronary angiogram: normal coronary arteries and left ventricular (LV) function.

on coronary angiography and risks involved. The diagnostic yield of myocardial biopsy has been reported to be low in clinical trials.²

She was readmitted 6 weeks later with recurrence of chest pain, temporal headaches, jaw claudication, scalp pain, worsening of the aches in the shoulder girdle and further weight loss. Clinical examination revealed tender temporal artery pulses, a triphasic pericardial friction rub and pericardial knock. ESR, CRP and white cell count was markedly elevated. Infection screen including paired-viral serology and auto-antibody screen including antinuclear antibodies, extractable-nuclear antigen, rheumatoid factor, antineutrophil cytoplasmic antibody were negative. Echocardiogram showed a small pericardial effusion, but normal LV function (figures 2 and 3). However, within a week, a repeat echocardiogram revealed a significant pericardial effusion over the right heart greater than the left without deterioration in cardiac function. A diagnosis of GCA related pericarditis was made on clinical grounds. She satisfied the American college of Rheumatology classification criteria for GCA (age more than 50 years, headache, temporal artery tenderness and ESR more than 50 mm in the first h).³ In addition, she had jaw claudication and scalp tenderness. A temporal artery biopsy (TAB) was considered

but not performed as the patient had been on prednisolone in the preceding 7 weeks and therefore, the likelihood of a negative result on TAB while on steroids.⁴ In addition, the clinical picture was quite characteristic of progression of PMR symptoms to frank GCA over the preceding months coupled with her previous presentation to the surgeons with unexplained normocytic anemia and weight loss. We would have proceeded to TAB if the clinical features had been atypical, but now agree with the subsequently published guidelines which make TAB mandatory.⁵ However, high dose of steroids (prednisolone 1 mg/kg) resulted in a dramatic resolution of symptoms again making GCA very likely. On subsequent inpatient and then outpatient review, the friction rub, pericardial knock and pericardial effusion had disappeared with again normal LV function on echocardiography (figures 2 and 3).

DISCUSSION

Patients presenting with chest pain can cause diagnostic dilemma especially when these pains are atypical with ECG changes. We have included a list of conditions with which patients present to acute care settings complaining of chest pain (table 1). This patient's ECGs showed dynamic ST elevation in the precordial leads and inferior and lateral saddle-

Table 1 Causes of chest pain without necessary rise in troponin

Cardiovascular	Gastroenterological	Respiratory	Musculoskeletal
Angina; Aortic dissection; Aortic stenosis; Arrhythmias; Myocarditis; Pericarditis; Valvular heart disease	Diverticular disease; Oesophageal spasm; Oesophageal rupture; Oesophagitis; Gastritis; Gastroesophageal reflux; Pancreatitis	Cancer; Chest trauma; Epiglottitis; Foreign Body; Pleurisy; Pneumothorax; Pneumomediastinum; Pneumonia; Pulmonary embolism; Tracheitis	Arthritis; Costochondritis; Fibromyalgia; Muscular strain; Trauma
Drug abuse	Psychological	Others	
Amphetamine; Cocaine; Diet pills; Ecstasy	Psycho-somatic disorder; Panic attacks; Depression	Anaemia; Abscess; Endocrine disorders; Infections – Herpes, Coxsackie	

shaped ST elevation rather than the usual global changes seen with pericarditis. There was also lower septum and apical marked hypokinesia with impairment of LV function suggestive of left coronary artery occlusion in addition to a small pericardial effusion. This patient had localised LV dysfunction suggestive of focal myocardial involvement either arising de novo or secondary to coronary arteritis but unusually without a rise in cardiac troponin T despite dynamic ST changes and localised LV dysfunction. With the dynamic ECG changes, an alternative diagnosis was myopericarditis. The patient was treated with steroids before coronary angiography a few days later, which may have improved a possible coronary arteritis in the left anterior descending artery territory and the normal appearances of the coronary arteries did not exclude a recent arteritis. Interestingly, the typical symptoms of temporal arteritis evolved many weeks after her presentation with chest pain.

Pericarditis is another complication which can rarely occur in GCA and fewer than 30 cases have been described in literature. Our case demonstrated the initial presentation with a small pericardial effusion with inferior and lateral saddle-shaped ST elevation on the ECG among other ECG changes. She also re-presented at a later date with typical features of GCA with further development of a larger pericardial effusion, but at the later presentation without LV dysfunction.

Involvement of branches of external carotid artery is common in GCA. Very rarely, aorta and other arteries are involved leading to complications in the affected arterial territory which include aortic aneurysm, dissection and rupture. One study had shown that the cranial symptoms (headache, scalp tenderness, abnormal temporal arteries) were negatively associated with large-artery stenosis as was a higher ESR.⁶

There have been a few case reports of fatal AMI⁷ and severe heart failure requiring cardiac transplantation.⁸ The incidence of fatal AMI due to GCA in the coronary arteries is extremely rare,⁹ as is non-fatal AMI.¹ AMI and severe heart failure have also occurred during treatment of GCA.⁹ Cardiac involvement including aorto-coronary disease in GCA can occur and manifest before other symptoms. The pathogenesis is unknown but postulated to be due to inflammatory cytokines activation leading to immunological damage. Immunoglobulin M (IgM) deposits have been found in coronary arteries.¹⁰ Mural thrombi formed during AMI can throw emboli to the cerebral circulation precipitating a cerebrovascular accident.¹¹

The diagnosis of coronary involvement should be suspected in patients who present with chest pain however atypical their symptoms may be. A high index of suspicion for GCA should be there for older patients who present with recurrent pericarditis-like symptoms as many patients

show a delayed onset of temporal arteritis and a TAB should be considered. The British Society for Rheumatology has recently recommended a TAB whenever GCA is suspected preferably within 1 week of starting steroids.⁵ An echocardiographic assessment is useful and coronary angiography may be necessary in selected cases.

Most patients can be managed with high dose steroids to suppress inflammatory activity; co morbidities including hypertension, diabetes mellitus and hyperlipidaemia which commonly occur in the older population may also need to be addressed. The use of antiplatelet agents and heparin at the time of initiation of steroids in coronary arteritis has been advocated by some workers,¹² but remains controversial. Patients should be monitored closely with clinical assessments and laboratory inflammatory markers. ESR and CRP usually settle in 2–4 weeks time in many cases. The true incidence of GCA complicating atherosclerotic coronary disease will always be difficult to define in the older population. As most cases are diagnosed retrospectively, the physician should be alert to the diverse presentations and complications of a systemic vasculitis like GCA so that early and appropriate treatment could be offered and thereby, reducing morbidity and mortality.

Learning points

- ▶ GCA is a systemic vasculitis with patchy vascular territorial localisation usually manifesting as temporal arteritis.
- ▶ The localisation in different vascular beds leads to heterogeneous manifestations and poses a considerable diagnostic challenge.
- ▶ Cardiac involvement is a rare complication of GCA.
- ▶ Coronary arteritis can occur before other common manifestations of GCA.
- ▶ Myopericarditis can mimic an AMI with pain and ECG changes which may be focal or generalised but when it presents with ECG changes in single arterial territory and LV dysfunction without increase in cardiac biomarkers such as troponin T, diagnosis becomes difficult and coronary angiography becomes necessary.
- ▶ Conditions mimicking AMI can cause diagnostic difficulties in the emergency room and may be initially treated with thrombolytic therapy or coronary angiography with primary percutaneous coronary intervention.
- ▶ A high index of suspicion is necessary and physicians should be alert to the rarer but significant complications of GCA as early treatment with steroids can reduce the likelihood of potentially severe outcomes.

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Competing interests None.

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