Case report

The nervous heart: a case report and discussion of an under-recognized clinical problem

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A 73-year-old Caucasian woman admitted to the neurology ward with Guillain–Barré syndrome (GBS) developed acute breathlessness 7 days after the onset of symptoms. Initial presentation was with symmetrical lower limb weakness, patchy sensory loss, absent tendon reflexes, normocellular cerebrospinal fluid (CSF) with raised protein (1.05 gm/l) and absent of waves on electromyography. There were no other associated cardiac symptoms, no neuromuscular respiratory weakness, normal neck muscle power, stable vital capacity and no hypercapnia on arterial blood gas analysis. She was a smoker. No arrhythmias were detected with continuous electrocardiogram (ECG) monitoring that is initiated on admission in the view of the well-recognized risk of autonomic dysfunction with GBS.

On examination, she was apyrexial, well perfused, in sinus rhythm at a rate of about 70 beats/min and normotensive. There was congestive cardiac failure with elevated jugular venous pressure, bibasal crepitations and a fourth heart sound. Her ECG (Figure 1b) demonstrated downsloping ST segments, T-wave inversion in inferior and precordial leads and a prolonged corrected QT interval at 490 ms. A portable transthoracic echocardiogram revealed apical and anteroseptal hypokinesia with impairment of left ventricular function. Troponin was mildly elevated (0.06 ng/ml, normal range <0.04 ng/ml). The differential diagnoses were Acute Coronary Syndrome (ACS) and stress cardiomyopathy. Atherothrombotic disease was a concern considering the prothrombotic risk with intravenous immunoglobulin. She was transferred to the coronary care unit and treated with dual antiplatelet therapy, low-molecular weight heparin and intravenous diuretics. Urgent coronary angiography revealed unobstructive coronary disease with no evidence of acute plaque rupture, excluding coronary disease as the cause of the ECG changes and acute ventricular dysfunction (Figure 1a). A follow-up echocardiogram performed at 6-month follow-up was normal. She made a good neurological recovery with mild residual distal leg weakness.

We made a diagnosis of stress cardiomyopathy, also known as Takotsubo Cardiomyopathy, rarely named Apical Ballooning Syndrome or Broken Heart Syndrome. The patient fulfilled the diagnostic criteria proposed by the Mayo Clinic (Table 1).

Discussion

Association of stress with cardiovascular events has been long recognized.2 Natural disasters, such as earthquakes, have allowed some insights into the influence of stress upon cardiac morbidity and mortality. The probability of short-term cardiac morbidity was increased by nearly 50% after an earthquake; and less than half of the sudden cardiac deaths in these studies could be explained by atherosclerotic disease.3 Similar findings have been reported in civilian populations during military conflict.4 Ventricular dysfunction in stress cardiomyopathy is reversible, and this clinical syndrome appears to represent a form of acute catecholamine-induced myocardial stunning.

Association of GBS with stress cardiomyopathy is not well understood. There have been reports...
of reversible cardiomyopathy in GBS which we believe represents the same disease spectrum. Supraphysiological serum catecholamine levels have been reported in several cohorts with acute stress cardiomyopathy, and appear central to the pathophysiology. This may explain the association with the acute phase of GBS. Dysregulation of autonomic tone with excessive sympathoadrenal activation in GBS has been reported with elevated catecholamines and their metabolites recorded in 24-h urine collection. Significant fluctuations in autonomic tone and high surges in serum catecholamine levels may be sufficient to induce acute catecholaminergic myocardial stunning in susceptible individuals. Our case is a postmenopausal woman, which appears to be one of the risk groups for stress

Figure 1. (a) Left coronary angiogram demonstrating no obstructive coronary disease. (b) ECG during the acute phase of illness showing ST segment changes and a prolonged QT interval.

Table 1 Diagnostic criteria proposed by Mayo Clinic for apical ballooning syndrome

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<td>Transient hypokinesis, akinesis or dyskinesis of the left ventricular midsegments with or without apical involvement; the regional wall motion abnormalities extend beyond a single epicardial vascular distribution; a stressful trigger is often, but not always present.</td>
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<td>Absence of obstructive coronary disease or angiographic evidence of acute plaque rupture.</td>
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<td>New electrocardiographic abnormalities (either ST-segment elevation and/or T-wave inversion) or modest elevation in cardiac troponin.</td>
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cardiomyopathy triggered by acute emotional or physical stress. It is interesting to note that other acute neuromuscular emergencies have also been reported to trigger stress cardiomyopathy. This reinforces the importance of considering cardiac causes of breathlessness in addition to progressive respiratory neuromuscular impairment and aspiration pneumonia in patients with acute progressive neuromuscular weakness. Histological and metabolic studies are in keeping with toxic catecholamine effects rather than ischaemic injury as the primary mediator.

The first description of stress cardiomyopathy with the characteristic left ventricular apical dysfunction was in 1981 from Japan. The authors proposed the name Takotsubo Cardiomyopathy after the traditional Japanese octopus trapping pot which has a characteristic long neck and a round bottom: a shape mimicked by the left ventriculogram in these patients. A preceding physical or emotional stressful event is found in about two-thirds of the patients.

It is important to consider stress cardiomyopathy in the differential diagnosis of ACS or acute left ventricular failure, especially in the setting of psychological or physiological stress. It is estimated that 0.52% of the patients presenting with clinical features of ACS have Stress Cardiomyopathy. There are increasing reports of stress cardiomyopathy in patients with severe and acute conditions, including the general intensive care population, a variety of non-cardiac medical emergencies. Latrogenic stress cardiomyopathy has been reported following electroconvulsive therapy, dobutamine stress echocardiography and intravenous epinephrine administration. The long-term cardiac prognosis in patients with stress cardiomyopathy is excellent. There is a significant acute mortality and morbidity in the small proportion of patients with Stress CM complicated by acute cardiogenic shock and secondary multi-organ failure.

The presentation can entirely mimic an ACS, with chest pain, breathlessness, ischaemic ECG changes including ST elevation or depression and QT prolongation. Fatal ventricular arrhythmias have been reported. Treatment targets acute left ventricular dysfunction with intravenous nitrates, diuretics and ACE inhibitors. Access to early diagnostic coronary angiography is critical to exclude coronary disease, and allow early cessation of ACS-based pharmacology. β-Blockers may improve short-term outcome, particularly in the setting of acute left ventricular outflow tract obstruction. Given the potential for an underlying phaeochromocytoma, β-blockers should be started cautiously in the younger patients, and those with atypical features, e.g. ‘Inverted Takotsubo Cardiomyopathy’. The long-term benefit of β-blockers, particularly in prevention of recurrence, remains to be shown. Anti-coagulation should be considered in patients with severe left ventricular dysfunction and is mandatory for patients with ventricular thrombus. Given the good long-term prognosis, patients with severe cardiogenic shock and deteriorating cardiac function should be treated ‘aggressively’, with intra-aortic balloon pump support, and left ventricular assist device implantation or extracorporeal life support may be warranted depending upon local availability. Inotropic drugs enhancing β-adrenoceptor signalling pathways are best avoided if possible.

Acute stress cardiomyopathy should be considered in the differential diagnosis of acute dyspnoea in patients with GBS, particularly if characterized by acute autonomic dysfunction. Early recognition of stress cardiomyopathy in GBS using echocardiography and coronary angiography is crucial in view of the excellent prognosis if complications are identified and addressed early. Incidence of this condition across other neurological emergencies remains largely unexplored and warrants further investigation.

Conflict of interest: None declared.

References