Ventricular tachycardia storm as predominant cardiac manifestation of lupus myocarditis

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SUMMARY

Systemic lupus erythematosus (SLE) is a multi-system autoimmune disease that can affect any part of the heart, causing arrhythmias on top of other cardiac manifestations. Malignant ventricular tachyarrhythmias are rare manifestations of SLE. Our case is the first one reported in the literature of an SLE patient with multi-organ involvement who subsequently presented with ventricular tachycardia (VT) storm as a cardiac manifestation. This case also demonstrates the use of Stellate ganglion block to treat intractable VT storm when chemical and electrical cardioversions failed, while waiting for immunosuppressive drugs to take effect. Timely treatment resulted in a good outcome for our patient.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a multi-system autoimmune disease that can affect any part of the heart, resulting in rhythm abnormalities presenting as either brady- or tachy-arrhythmias.[1] Malignant ventricular tachyarrhythmias are rare manifestations of SLE. We describe a case of SLE with multi-organ involvement who developed ventricular tachycardia storm as the main cardiac manifestation of her autoimmune disease. The clinical features, investigations, and management will be discussed.

CASE REPORT

A 50-year-old Malaysian-born Chinese lady presented to a private hospital in Sarawak, Malaysia with a 3-day history of chest discomfort, generalized myalgia, and lethargy. She was diagnosed with systemic lupus erythematosus (SLE) 4 years ago with multi-organ involvement including joints, eyes, mucosa, and muscle. She was prescribed oral hydroxychloroquine 200 mg OD, cyclosporine 50 mg BD, and mycophenolate mofetil 250 mg BD by her rheumatologist. Unfortunately, she was not compliant to her prescribed medications and had been taking traditional Chinese medicine instead for a month prior to this presentation.

She was a housewife who was previously a non-smoker, and a teetotaler. She did not have any family history of autoimmune disease.

Physical examination during her current admission was unremarkable. There was neither any sign of heart failure nor any mucocutaneous manifestation of active SLE. Blood investigations showed markedly elevated Troponin T of 1400 pg/mL, creatine kinase (CK) of 7723 U/L, creatine kinase-myoglobin binding (CK-MB) of 212 U/L, and brain natriuretic peptide (pro-BNP) of 1074 pg/mL. However, there was no electrolyte imbalance (Table I). Initial 12-lead electrocardiogram showed idioventricular rhythm (Figure 1) but she later developed pulseless ventricular tachycardia of 150–160 beats per minute which was negative in leads II, III, and aVF (Figures 2 and 3). Electrical cardioversion was delivered during the onset of pulseless VT with restoration of spontaneous circulation. A cardiologist input was immediately sought, and she was commenced on IV amiodarone with a loading dose of 300 mg over 1 hour followed by a maintenance dose of 900 mg over 23 hours. Subsequent transthoracic echocardiogram showed an LV ejection fraction of 50% with hypokinetic infero-septal wall. However, invasive coronary angiography revealed normal coronary arteries. Computed tomography of pulmonary angiography (CTPA) revealed no evidence of pulmonary embolism.

She was treated for lupus myocarditis and started on oral hydroxychloroquine 400 mg OD and IV methylprednisolone 500 mg OD for 3 days by the rheumatologist. Concurrent oral metoprolol was given. IV cyclophosphamide 500 mg was given on day 2 of admission to keep her lupus myocarditis under control.

On the third day, she developed recurrent unstable monomorphic VT that required 5 boluses of 150 mg IV amiodarone (total dose 750 mg). A diagnosis of VT storm secondary to lupus myocarditis was made. IV immunoglobulin 70 g was commenced at 10 g/hour over 7 hours for 2 days. On the fourth day, she developed transient thyroid dysfunction with raised free T4 but amiodarone was continued after consultation with the endocrinologist. Later in the evening, she again developed recurrent VT requiring repeated boluses of IV metoprolol up to 9 mg in total (2 mg, 2 mg, and 5 mg). She was subsequently intubated and sedated to suppress sympathetic activity. Deep sedation was achieved with IV propofol, fentanyl, and midazolam aiming for a target Richmond Agitation-Sedation Scale of -4 (deep sedation) to -5 (unarousable sedation).

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Table I: Laboratory test results on the day of presentation

Blood test	Results	Reference range
Haemoglobin level	13.9	12.0–15.0 g/dL
White cell count	4.22	4.0–10.0 x 10^3
Platelet count	251	150–410 x 10^9
Sodium	142	135–145 mmol/L
Potassium	3.3	3.3–5.1 mmol/L
Urea	18.2	1.7–8.3 mmol/L
Creatinine	111	50–98 mmol/L
Calcium (corrected)	2.09	2.20–2.60 mmol/L
Phosphate	1.95	0.74–1.52 mmol/L
Magnesium	2.10	0.66–1.07 mmol/L
Albumin	29	35–50 g/L
Aspartate aminotransferase	441	1–37 U/L
Alanine aminotransferase	605	1–40 U/L
high sensitive Troponin T	590.5	0–14 pg/mL
NT-proBNP	1510	<125 pg/mL (patients aged 0-74 years), and <450 pg/mL (patients aged 75-99 years)

Her oral beta-blocker was changed to IV metoprolol infusion at 3 mg/hour on the fifth day of admission. IV hydrocortisone 50 mg QID was commenced after 3 days of IV methylprednisolone.

She developed fever 4 days into admission but there was no obvious source of infection. Covid-19 PCRs taken on the day of admission and at fever onset were both negative and white blood cell count was not raised (4.22 x 10^3; reference range 4–10 x 10^3). Blood cultures on admission showed no growth. Nevertheless, she was started on IV Imipenem to cover for occult sepsis.

Despite treatment with IV amiodarone, IV metoprolol, and immunosuppressive drugs, she developed incessant unstable VT requiring 22 cycles of electrical cardioversion on the fifth day of admission. She was then transferred to the cardiology team at Sarawak Heart Center for left Stellate ganglion block. During transfer, she developed three further episodes of VT in the ambulance and one episode upon arrival at the cardiac intensive care unit, which required four more synchronized electrical cardioversion. Ultrasound-guided stellate ganglion block (SGB) with 5 ml of 2% lignocaine was done. The IV amiodarone was terminated after 24 hours due to a prolonged QT interval of 624 ms. IV metoprolol was changed to oral carvedilol 25 mg BD. There was no further VT episode after SGB as the immunosuppressive drugs started to take effect.

She was extubated after 3 days. Post extubation, there were copious amounts of respiratory secretions. Mucolytic and normal saline nebulisation were given. A day after extubation, she developed acute respiratory acidosis and carbon dioxide narcosis with Glasgow Coma Scale of 5 which necessitated re-intubation. CT scan of the brain did not show any intracranial bleed or infarction. Two days after the second intubation, she was extubated again. However, her respiratory effort was poor despite aggressive chest physiotherapy requiring noninvasive ventilation (NIV) for a day. Examination revealed poor gag reflex, weak respiratory effort, and proximal myopathy. Despite NIV, arterial blood gas analysis showed worsening acute respiratory acidosis. A third intubation was decided due to poor respiratory effort and carbon dioxide retention secondary to critical illness myopathy.

IV amoxicillin-clavulanate was given for 3 days after 3 days of IV meropenem. In view of clinical deterioration, her antibiotic treatment was then escalated to IV piperacillin-tazobactam for 4 days before de-escalation to IV amoxicillin-clavulanate again. Multiple blood cultures yielded no growth.

After a few days of immunosuppression, Troponin T and CK levels improved from initial values of 1400 pg/mL and 7723 U/L to 476.8 pg/mL and 397 U/L, respectively. N-terminal pro-brain natriuretic peptide (NT-proBNP) levels also improved from an initial value of 1510 pg/mL at presentation to 449 pg/mL.

Transfer of care to the rheumatology team at Sarawak General Hospital was done for rehabilitation in view of general deconditioning due to critical illness for which she required tracheostomy and Ryle's tube feeding. Her stay was further complicated with COVID-19 infection, Stage 2 pressure sore, and deep vein thrombosis of her left calf. Immunosuppression with fortnightly IV cyclophosphamide 500 mg during her stay had resulted in the remission of the patient's disease. There was still residual myopathy after 2 months of physiotherapy and rehabilitation whereby she was unable to mobilize and needed assistance to sit-up. However, her general condition gradually improved, and she was well enough to be discharged for basic home care by family members after a total of 3 months hospital stay. Cardiac magnetic resonance imaging (CMRI) done 3 months after treatment showed no signs of myocardial inflammation or scarring. She is now ambulating without support and continues to do well with oral prednisolone, hydroxychloroquine, and mycophenolate mofetil.

DISCUSSION

Lupus myocarditis is a rare presentation of SLE flare.[2] VT as a manifestation of lupus myocarditis is not a commonly encountered clinical scenario. VT storm as a cardiac manifestation of the autoimmune disease is even rarer. Our case is the first reported case of VT storm as a cardiac manifestation of SLE. VT storm is defined as three or more episodes of sustained VT within 24 hours necessitating abortion by an intervention.[3] Factors that increase the susceptibility of development of VT storm include male sex, advanced age, heart failure, and underlying comorbidities.[4]

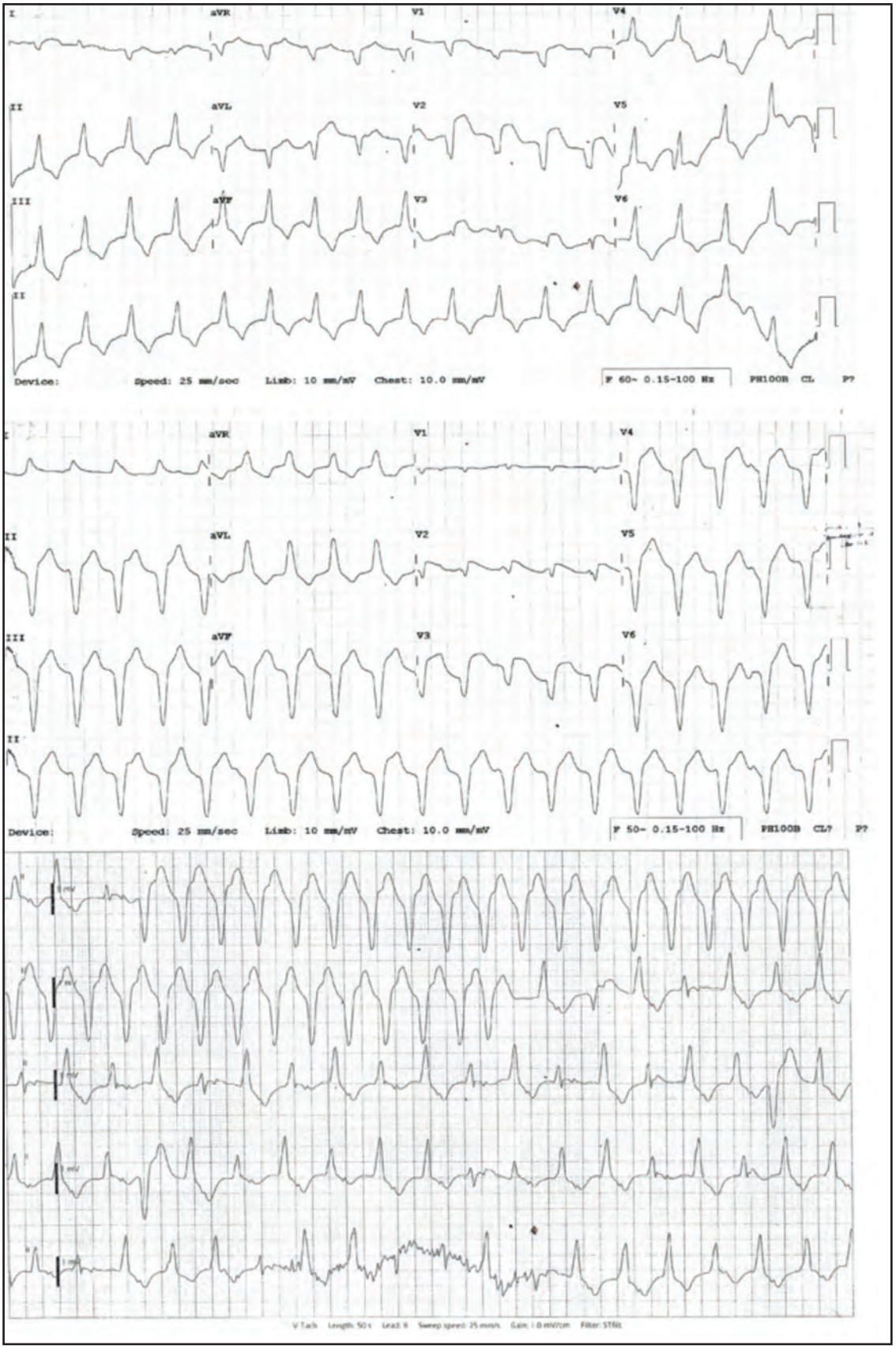

Fig. 1: Electrocardiogram of the patient. (A) A 12-lead electrocardiography of accelerated idioventricular rhythm. (B) 12-lead electrocardiography during the development of pulseless ventricular tachycardia. (C) 12-lead electrocardiography during one of the VT episodes with negative axis in II, III, aVF leads.

For our patient, the main factor was her underlying autoimmune disease.

The development of VT storm is due to a complex interplay of the triad of susceptible electrophysiological substrates, triggers, and autonomic dysregulation.[5] Neural remodeling caused by many chronic cardiac conditions causes parasympathetic input decrement and hyperinnervation of the sympathetic system leading to an increment of sympathetic activity that triggers the arrhythmia.[6]

Anti-arrhythmic agents are important in the treatment of arrhythmias. Amiodarone was chosen for our patient as the initial drug. It is a useful drug for the treatment of arrhythmias, but its adverse effects are vast including corneal deposits, photosensitivity, hepatitis, and thyroid dysfunction.[7] Our patient experienced thyroid function derangement from the treatment. Deep sedation and mechanical ventilation should be considered in intractable VT storm after adequate anti-arrhythmic agents to counteract the sympathetic hyperactivity, aiming for Richmond Agitation-Sedation Scale values below -2.[8]

Autonomic modulation has been described by many studies as a treatment for VT storm.[6,9] Percutaneous stellate ganglion block (SGB) is one of the techniques to achieve autonomic modulation. It can be done as a bedside procedure under ultrasound guidance by injecting local anaesthetic into the left stellate ganglion or bilateral stellate ganglia. The stellate ganglion is located behind the carotid artery. Hence, the procedure involves the advancement of needle in a lateral-to-medial direction to the anterior surface of the longus coli muscle to avoid injury to major blood vessels. Local anaesthetic is injected after a negative aspiration. Our choice of local anaesthetic for our patient was 5 ml of 2% lignocaine. Our case demonstrated that SGB can be an effective temporary measure after anti-arrhythmic therapy while waiting for immunosuppression to take its effect in the setting of VT storm in lupus myocarditis.

An implantable cardioverter defibrillator (ICD) can be considered for secondary prevention after CMRI to look for any structural abnormality or ventricular scarring as the potential pathological causes of the VT storm. CMRI can also diagnose myocarditis. For our patient, due to the requirement of emergency resuscitation at the time of presentation, diagnosis of myocarditis was made based purely on clinical grounds and raised Troponin T without radiological evidence. CMRI done 3 months after treatment showed no sign of myocardial inflammation or scarring. This might indicate the effectiveness of our timely administration of immune-modulatory therapy. An ICD implant for secondary prevention was not indicated due to the normal CMRI findings.

CONCLUSION

Lupus myocarditis with VT storm is a rare manifestation of SLE flare. Anti-arrhythmic drugs are useful in the initial treatment of VT storm. Deep sedation and mechanical ventilation are the next steps in the management of intractable VT. Immunosuppression should be initiated to treat lupus myocarditis. However, while waiting for immunosuppression to take effect, SGB can be an effective temporary measure to treat intractable VT storm when other therapies have failed. Our case illustrated the successful use of SGB for the treatment of VT storm in lupus myocarditis.

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CONFLICT OF INTEREST

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