Dear Editor,

A 43-year-old Myanmese woman with a background history of systemic lupus erythematosus (SLE) presented with a 1 week history of lethargy, headache, exertional dyspnoea and near syncope a week after her 2nd dose of SARS-CoV2 BNT162b2 mRNA vaccination. She had a few days of gum bleeding and bruising over her left wrist. She reported no alopecia, rash, ulcers or joint pains, which were her initial manifestations of SLE, first diagnosed in 2008. Her predominant disease activity was marked by intermittent immune thrombocytopenia. Her disease had been in remission on azathioprine, hydroxychloroquine and prednisolone with no flares for the last 2 years. She had been compliant to her medication, with no traditional medicines or supplements. On examination, she was pale and lethargic looking, with no photosensitive rash, alopecia or oral ulcers. Cardiovascular and neurological examinations were unremarkable.

Her blood counts showed macrocytic anaemia (haemoglobin level 5.8 g/dL, MCV 107 fL) with severe thrombocytopenia (platelet count $7 \times 10^9$/L). There was evidence of haemolysis with a positive direct Coombs test, haptoglobin <30 mg/dL and a raised LDH of 2226 U/L. Complements were low (complement3 0.64 g/L, complement4 0.08 g/L). Reticulocytes were elevated $125.9 \times 10^9$/L. Renal function and electrolytes were normal. Hepatitis B, hepatitis C and human immunodeficiency virus serologies were negative. Electrocardiogram showed new left bundle branch block. Troponin I was mildly elevated, 48 ng/L (0–18 ng/L) with ejection fraction 50% and mild global hypokinesia on echocardiography. Computed tomography of the brain showed no intracranial bleed, and chest x-ray showed no consolidation or pleural effusion.

The new-onset autoimmune haemolytic anaemia and immune thrombocytopenia were consistent with Evans syndrome (ES), which signified a major SLE flare, in the setting of hypocomplementaemia, high-titre anti-dsDNA and possible myocarditis.

She received pulse intravenous methylprednisolone and was subsequently commenced on intravenous immune globulin and rituximab due to slow-rising platelet and haemoglobin counts. Her platelet counts gradually improved to $231 \times 10^9$/L after 2 weeks. Her haemoglobin rose to 10.6 g/dL (11.8–14.6 g/dL), and LDH levels improved from 2226 to 546 U/L, with normalization of complements and declining anti-dsDNA.

SARS-CoV2 mRNA vaccines have been shown to be effective in preventing COVID-19 infection. However, the vaccine has been associated with flares of autoimmune haematological disorders [1–3] and autoimmune conditions like SLE [4]. In this patient, the SARS-CoV2 vaccine had triggered an SLE flare with a new manifestation of ES, in an individual whose disease activity was previously only marked by intermittent thrombocytopenia and was under control with immunosuppressants.

The underlying pathophysiology of ES is not well understood but can be primary or secondary to autoimmune haematological disorders [1–3] and autoimmune conditions like SLE [4]. In this patient, the SARS-CoV2 vaccine had triggered an SLE flare with a new manifestation of ES, in an individual whose disease activity was previously only marked by intermittent thrombocytopenia and was under control with immunosuppressants.

The underlying pathophysiology of ES is not well understood but can be primary or secondary to autoimmune conditions or viral infections [5]. SLE is also the most frequent autoimmune disease associated with ES [6].

Currently, patients with autoimmune and rheumatic conditions are still recommended to receive the mRNA vaccine over other forms of vaccination with an mRNA vaccine as a supplemental dose as the expected benefits of vaccination would...

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**LETTER TO THE EDITOR**

**New-onset Evans syndrome in a patient with SLE post SARS-CoV2 mRNA vaccination**

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are thought to outweigh the risk of disease flare [7]. Close monitoring in patients who have underlying autoimmune conditions is recommended.

Declarations

Ethical approval  This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent  Informed consent was obtained from all individual participants included in the study.

Disclaimer  The patient has consented to the use of relevant information for academic purposes.

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