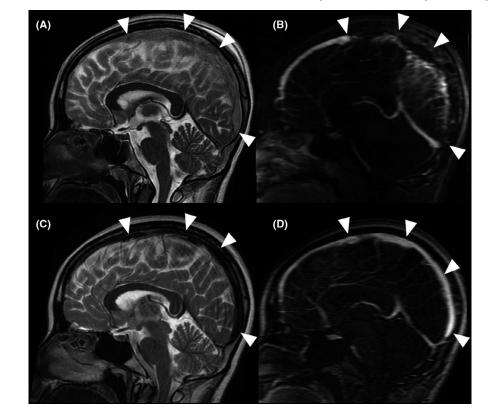
ACR Open Rheumatology

Vol. 3, No. 6, June 2021, pp 395 © 2021 The Authors. ACR Open Rheumatology published by Wiley Periodicals LLC on behalf of American College of Rheumatology. This is an open access article under the terms of the Creative Commons Attribution-NonCommercial License, which permits use, distribution and reproduction in any medium, provided the original work is properly cited and is not used for commercial purposes.

DOI 10.1002/acr2.11262



Clinical Images: Cerebral venous sinus thrombosis as one of the initial presentations of systemic lupus erythematous

The patient, an 18-year-old woman, presented to our hospital with skin rash, nausea, vomiting, and headache for 1 week. The physical examination revealed a malar rash. The laboratory test results showed pancytopenia, proteinuria, and high levels of antinuclear and anti-doublestranded DNA antibodies. The diagnosis of systemic lupus erythematosus (SLE) was established. Hydroxychloroquine and methylprednisolone were prescribed. However, the headache deteriorated, and consciousness became drowsy. The sagittal T2-weighted brain magnetic resonance imaging showed dilated superior sagittal sinus with heterogeneously high signal (A, arrowheads). The contrast-enhanced magnetic resonance venography at the corresponding location showed a segmental filling defect spanning from the middle superior sagittal sinus to the confluence of sinuses (B, arrowheads). These findings suggested the diagnosis of cerebral venous sinus thrombosis. Relevant laboratory test results included a mildly elevated level of anticardiolipin immunoglobulin G (10 U/ml; negative, less than 10 U/ml), a positive lupus anticoagulant test result (ratio of 1.28; negative, ratio of less than 1.2), and an elevated level of d-dimer (21.5 mg/l; negative, less than 0.5 mg/l). Low-molecular-weight heparin, medium-dose methylprednisolone, and cyclophosphamide were administered. She had an excellent clinical response, without sequelae. After 6 months of treatment, follow-up brain magnetic resonance imaging showed resolution of thrombosis (C and D, arrowheads). Cerebral venous sinus thrombosis is a rare complication of SLE (1). Headache, visual field defects, and altered consciousness are the leading presentations of cerebral venous sinus thrombosis in SLE (1). Delayed diagnosis is associated with an increased risk of later visual deficit (2). In addition, venous hypertension due to venous thrombosis may cause venous infarction, cerebral edema, intracranial hypertension, cerebral hernia, and death (1). Intracranial hemorrhage related to cerebral venous sinus thrombosis was also reported to be associated with mortality (3). Therefore, early diagnosis of cerebral venous sinus thrombosis and immediate anticoagulation are essential for these patients.

- Duman T, Demirci S, Uluduz D, Kozak HH, Demir S, Misirli CH, et al. Cerebral venous sinus thrombosis as a rare complication of systemic lupus erythematosus: subgroup analysis of the VENOST study. J Stroke Cerebrovasc Dis 2019;28:104372.
- Saposnik G, Barinagarrementeria F, Brown RD Jr, Bushnell CD, Cucchiara B, Cushman M, et al. Diagnosis and management of cerebral venous thrombosis: a statement for healthcare professionals from the American Heart Association/American Stroke Association. Stroke 2011;42:1158–92.
- 3. Luo Y, Tian X, Wang X. Diagnosis and treatment of cerebral venous thrombosis: a review. Front Aging Neurosci 2018;10:2.

Chiao-Feng Cheng, MD D National Taiwan University Hospital Yunlin Branch, Yunlin County, Taiwan Ko-Jen Li, MD, PhD D National Taiwan University Hospital Taipei, Taiwan