Comment on: Reversible cerebral vasoconstriction syndrome in a female patient with systemic lupus erythematosus

Sir, Sayegh and colleagues [1] reported reversible cerebral vasoconstriction syndrome (RCVS) in a female patient with SLE. This appears to be the first case of RCVS in literature described in an SLE patient. However, we wonder if the authors have considered differential diagnoses other than lupus-associated cerebral vasculitis, i.e. subarachnoid haemorrhage (SAH).

The clinical manifestations of the patient, including acute severe headache associated with nausea and vomiting, as well as haematic cerebrospinal fluid, are strongly suggestive of a diagnosis of SAH. SAH can occur in SLE regardless of disease activity [2]. Although the head CT scan was normal for this case, negative CT findings in SAH are not uncommon [3]. RCVS is a heterogeneous entity, characterized by severe headaches and constriction of cerebral arteries that resolves spontaneously in 1–3 months [4]. The pathophysiology of RCVS remains unknown, while the prevailing hypothesis is that a transient disturbance in the control of cerebral vascular tone leads to segmental and multifocal arterial constriction and dilatation [5]. Since RCVS can be either preceded or succeeded by SAH [4], diagnosis of ambiguous RCVS may not be as clinically important as SAH per se with regard to treatment. We agree that multifocal segmental stenosis of cerebral arteries can be seen from magnetic resonance angiography (MRA) [1]. However, CNS-SLE (headaches, psychosis and seizure) and poor treatment adherence may hardly exclude the diagnosis of lupus-associated cerebral vasculitis, which might remit after CYC and prednisone treatment. Another assumption is that vasospasm, possibly resulting from SAH [6], recovered when haemorrhage subsided. Multiple lines of evidence other than severe headache and haematic cerebrospinal fluid point to the occurrence of SAH in this patient. Lupus-associated cerebral vasculitis [2], or disorders in blood coagulation system either resulting from SAH per se, or from utilization of prednisone or atorvastatin [7], rank as risk factors for SAH in this case.

Taken together, the complex clinical manifestations of SLE pose a great challenge to clinicians in differentiating headache. Analysis of blood coagulation function and blood cells, repeated CT examination and physical examination for nuchal rigidity, might be of help to exclude SAH and initiate corresponding treatment in patients with thunderclap headache.

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References


Letters to the Editor

Comment on: Reversible cerebral vasoconstriction syndrome in a female patient with systemic lupus erythematosus: reply

Sir, We would like to thank Zhang et al. [1] for their interest and comments on our manuscript reporting a case of reversible cerebral vasoconstriction syndrome (RCVS) in a female SLE patient [2]. Zhang et al. [1] raise the hypothesis that the clinical manifestations of the patient were due to subarachnoid haemorrhage (SAH) rather than RCVS. We would like to make the following comments.

RCVS is a clinical–radiological entity characterized by the association of recurrent acute ‘thunderclap’ head...