Histiocytic necrotising lymphadenitis identical to Kikuchi-Fujimoto disease in CNS lupus

Maria Ioanna Stefanou,[1] German Ott,[2] Ulf Ziemann,[1] Annerose Mengel[1]

[1]Department of Neurology and Stroke, and Hertie Institute for Clinical Brain Research, University of Tübingen, Tübingen, Germany
[2]Department of Clinical Pathology, Robert-Bosch Krankenhaus, and Margarete Fischer-Bosch Institute for Clinical Pharmacology, Stuttgart, Germany

Correspondence to
Dr Maria Ioanna Stefanou, maria-ioanna.stefanou@med.uni-tuebingen.de

Accepted 25 May 2018

DESCRIPTION

A 36-year-old woman presented after a first generalised tonic–clonic seizure. On clinical examination, prominent cervical and axillary lymphadenopathy was noted. Brain MRI showed diffuse, partially enhancing, hyperintense lesions (figure 1). An axillary lymph node biopsy revealed typical findings of histiocytic necrotising lymphadenitis, compatible with Kikuchi-Fujimoto disease (figure 2).[1] Laboratory studies showed positive antinuclear antibodies, low complement levels and pancytopenia. In the cerebrospinal fluid (CSF), lymphocytic pleocytosis (34/μL), elevated lactate (3.3 mmol/L) and protein (104 mg/dL) levels were noted. Investigations for lymphoma were negative. Furthermore, negative serological and PCR findings for Epstein-Barr virus, cytomegalovirus and HIV were found. Interferon-gamma release assay and PCR for tuberculosis, along with serological investigations for toxoplasmosis and syphilis, were negative. The cytological examination of the CSF revealed activated lymphocytes and plasma cells. CSF and blood cultures remained negative. On diagnostic evidence of pericarditis and a positive history for photosensitivity and arthralgia, the diagnosis of a central nervous system (CNS) manifestation of systemic lupus erythematosus was made.[2] Two months after intense immunosuppressive therapy with methylprednisolone, rituximab and cyclophosphamide, the patient's clinical and paraclinical findings improved markedly.

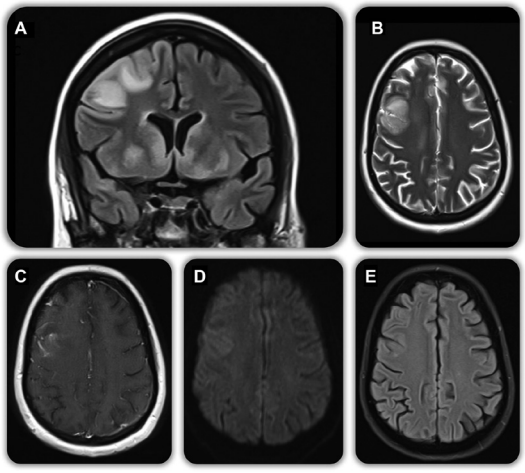

Figure 1 Brain MRI. Coronal Fluid-attenuated inversion recovery (FLAIR) (A) and axial T2w (B) MRI reveal cortical and subcortical hyperintensities involving the right frontal, temporal and parietal lobes, and the bilateral basal ganglia. Axial gadolinium-enhanced T1w MRI (C) demonstrates contrast enhancement without diffusion restriction (D). Intense immunosuppressive therapy resulted in significant remission of the MRI abnormalities (E).

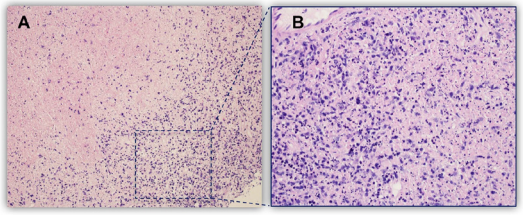

Figure 2 Lymph node biopsy specimen. At low magnification (A), extensive necrosis, surrounded by partly preserved lymphatic tissue with reactive changes, was noted (H&E, ×100). Higher magnification (B) showed extensive pyknotic and karyorrhectic nuclear debris, along with histiocytic cells and activated lymphocytes (H&E, ×400). The pathological findings were typical for histiocytic necrotising lymphadenitis, morphologically identical to Kikuchi-Fujimoto disease.

Kikuchi-Fujimoto disease, or histiocytic necrotising lymphadenitis, is a rare, benign and self-limited disease, affecting mainly young women and presenting with localised lymphadenopathy, fever and leucopenia in the majority of the reported cases.[1] The aetiology of Kikuchi-Fujimoto disease remains unknown. The hallmark histopathological feature is a histiocytic necrotising lymphadenitis, characterised by necrotic and/or histiocytic cellular infiltrates within the lymph node, often accompanied by invasion of the node capsule and inflammation of the perinodal tissue. The present report illustrates that histiocytic necrotising lymphadenitis, morphologically identical to Kikuchi-Fujimoto disease, may occur in association with systemic lupus erythematosus. As no effective treatment has been established for Kikuchi-Fujimoto disease, clinical awareness of the overlapping features of the two disorders is crucial for the prompt diagnosis and early initiation of immunosuppressive therapy in patients with concomitant systemic lupus erythematosus.

Learning points

- Kikuchi-Fujimoto disease, or histiocytic necrotising lymphadenitis, is a rare, benign and self-limited disorder of unknown origin.
- Kikuchi-Fujimoto disease may occur in association with systemic lupus erythematosus.
- High clinical awareness is warranted for early diagnosis and treatment initiation in patients with concomitant systemic lupus erythematosus, presenting with histiocytic necrotising lymphadenitis.

To cite: Stefanou MI, Ott G, Ziemann U, *et al*. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2018-225668

Acknowledgements The authors would like to acknowledge the significant contribution of Dr Hennersdorf in reporting the imaging studies.

Contributors MIS: case report conception, organisation and execution; writing of the first draft, review and finalisation, treating physician. GO: conduction and analysis of the pathological study, critical review of the manuscript. UZ: supervision, critical review and finalisation of the manuscript. AM: case report conception, organisation and execution, treating physician and coordinator of the project.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 Mahajan T, Merriman RC, Stone MJ. Kikuchi-Fujimoto disease (histiocytic necrotizing lymphadenitis): report of a case with other autoimmune manifestations. *Proc* 2007;20:149–51.
2 Smith EL, Shmerling RH. The American College of Rheumatology criteria for the classification of systemic lupus erythematosus: strengths, weaknesses, and opportunities for improvement. *Lupus* 1999;8:586–95.