Letters to the Editor

Lymphadenopathy Syndrome in Systemic Lupus Erythematous: Is It Kikuchi-Fujimoto Disease?1

Síndrome poliadenopático en lupus eritematoso sistémico: ¿Es la enfermedad de Kikuchi-Fujimoto?

To the Editor,

Lymphadenopathy syndrome is a common manifestation of systemic lupus erythematosus (SLE). In general, enlarged lymph nodes are small and can be found in the cervical, inguinal and axillary regions. Lymph node involvement is present in up to 25% of the patients and normally appears in the first stages of the disease or during relapses.1 The differential diagnosis of lymphadenopathy syndrome in SLE includes histiocytic necrotizing lymphadenitis or Kikuchi-Fujimoto disease (KFD), Castleman’s disease, syphilis, tuberculosis, sarcoidosis, infectious mononucleosis (Epstein–Barr virus [EBV], cytomegalovirus), herpes simplex, human immunodeficiency virus (HIV), hepatitis B virus (HBV), hepatitis C virus (HCV), other infections and lymphoma.2

Kikuchi-Fujimoto disease, or histiocytic necrotizing lymphadenitis, is a rare clinical disorder characterized by lymph node involvement (generally cervical), fever, night sweats and leukopenia. It mainly affects young Asian women. It was first described by Kikuchi and Fujimoto in 1972.3,4 The associated mortality is increased by heart, lung and liver involvement. The exact cause of KFD is not yet known, but recent publications are inclined toward a viral infection (EBV and others) or an autoimmune disorder (an exaggerated immune response mediated by T cells).5 To diagnosis the disease, it is necessary to perform an excisional biopsy of the affected lymph nodes. The association between KFD and SLE is rare, and whether it is incidental or a clinical manifestation of SLE continues to be a matter of debate.

The coexistence of these 2 diseases is reported in 2 young SLE patients (cases nos. 1 and 2) and we describe another case of lymphadenopathy syndrome in SLE (case no. 3) in which the affected lymph node was studied by cytology rather than excisional biopsy (Table 1). In all 3 cases, the results of serological testing for HIV, HBV, HCV, EBV, cytomegalovirus, herpes simplex virus, rubella, toxoplasma, parvovirus B19, Yersinia enterocolitica, Salmonella and Brucella were negative, as were the results of interferon-γ release assays (IGRA) for all 3 patients.

Kikuchi-Fujimoto disease has been associated with SLE and other connective tissue diseases, such as antiphospholipid syndrome, Sjögren’s syndrome, relapsing polychondritis and autoimmune hepatitis.6,7 It is generally a benign disease that resolves spontaneously in 1–4 months, but cases with a poor outcome and

Table 1
Description of the Three Cases.

<table>
<thead>
<tr>
<th>Case no. 1</th>
<th>Case no. 2</th>
<th>Case no. 3</th>
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</thead>
<tbody>
<tr>
<td><strong>Demographic</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age (years)</td>
<td>36</td>
<td>30</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>Female</td>
</tr>
<tr>
<td>Race</td>
<td>Caucasian</td>
<td>Latin American</td>
</tr>
<tr>
<td><strong>Clinical manifestations</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cervical lymph node involvement</td>
<td>Arthritis</td>
<td>Axillary and supraclavicular lymph node involvement</td>
</tr>
<tr>
<td>Asthenia</td>
<td></td>
<td>Polyarthritis</td>
</tr>
<tr>
<td><strong>Laboratory</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ANA</td>
<td>1/320</td>
<td>ANA 1/320</td>
</tr>
<tr>
<td>Anti-dsDNA</td>
<td>417 IU/mL</td>
<td>Anti dsDNA: 310 IU/mL</td>
</tr>
<tr>
<td>C3</td>
<td>75 mg/dL</td>
<td>C3: 112 mg/dL</td>
</tr>
<tr>
<td>C4</td>
<td>12 mg/dL</td>
<td>C4: 24 mg/dL</td>
</tr>
<tr>
<td><strong>Image</strong></td>
<td></td>
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</tr>
<tr>
<td>Cervical CT</td>
<td>Thoracoabdominal CT</td>
<td>Cervical CT</td>
</tr>
<tr>
<td><strong>Pathology</strong></td>
<td></td>
<td></td>
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<tr>
<td>Cervical lymph node: lymphocytes with areas of necrosis at different stages of maturation</td>
<td>Supraclavicular lymph node: lymphocytes with areas of necrosis at different stages of maturation</td>
<td>Excisional biopsy not performed</td>
</tr>
<tr>
<td>Numerous CD8-positive lymphocytes with TIA1+ cytotoxic granules (Fig. 1A–C)</td>
<td>Numerous CD8-positive lymphocytes with TIA1+ cytotoxic granules (Fig. 1D)</td>
<td>Malignancy ruled out using ultrasound-guided FNAC</td>
</tr>
<tr>
<td><strong>Treatment and outcome</strong></td>
<td>Glucocorticoids</td>
<td>Glucocorticoids</td>
</tr>
<tr>
<td>Lymph node involvement almost completely disappeared</td>
<td>Azathioprine</td>
<td>Lymph node involvement almost completely disappeared</td>
</tr>
</tbody>
</table>

ANA, antinuclear antibodies; CT, computed tomography; dsDNA, double-stranded DNA; FNAC, fine-needle aspiration cytology.

Biopsy

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56

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There

(2)

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Rheumatol


References


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http://dx.doi.org/10.1016/j.reumae.2016.04.004

2173-5743/

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