in the light zone of ectopic GC expressing CXCL13. One pSS in HTLV-I carrier showed a relatively small size of ectopic GC whose CXCL13 expression pattern was similar. However, interestingly, the MNCs of HAM-pSS patients demonstrated no expression of CXCL13. In contrast to CXCL13, CXCL12 was commonly expressed on ductal epithelial cells of all the pSS patients irrespective of anti-HTLV-1 antibody. In a normal subject, no expression of CXCL13 was observed with positive expression of CXCL12 similar with pSS.

The lymphoid aggregates of LSGs are responsible for autoimmune production that locally occurs in ectopic GC. Radiographic destruction of the ductal structure in HTLV-I-seropositive pSS occurs to a lesser extent than in HTLV-I-seronegative pSS, which is a unique characteristic of the former [4].

The chemokines have been found to regulate ectopic GC formation of SS [5]. Xanthou et al. [9] also demonstrated the significance of lymphoid chemokines for lymphoid structure formation in SS, while others have demonstrated an association of CXCL13 expression and ectopic GC formation in SS [7,8]. Barone et al. [8] found a B cell-dominant expression pattern, whereas the selected expression in acinar and ductal epithelial cells was observed by Salomonsson et al. [7], although the exact roles of these results remain unclear.

Our data suggest an important interaction of CXCL13 and ectopic GC in sialadenitis in SS. The tendency towards low levels of radiographic damage in patients with HAM-pSS suggests that salivary-specific cytotoxicity is modified by HTLV-I infection. Due to even expression of CXCL12 irrespective of HTLV-I infection, HTLV-I presumably affects the CXCL13 expression of infected CD4+ T cells. Via inflammatory mediators modulated by HTLV-I tax protein, dysfunction of MNC-lineage cells due to HTLV-I infection is supposed to play an important role.

**Rheumatology key message**

- Low prevalence of ectopic GC is a characteristic of HTLV-I-associated SS with CXCL13 on MNCs.

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in employment. Patient A had resided in rural South Australia for 39 years prior to the biopsy, whereas Patient B had always lived in urban areas. An infectious agent triggering IIM has not been isolated. The report of DM and SLE in monozygotic twin pairs [3] suggests a genetic predisposition to autoimmune disease, the phenotype being modulated by as yet undefined (environmental) factor. In IIM, further support for a role of non-infectious environmental factors in modulating disease expression is the influence of geographic latitude [4] and ultraviolet irradiance [5] on the relative prevalence of DM and PM.

Secondly, the validity of Patient B’s diagnosis needs questioning (was this undiagnosed IBM?), as emerging literature questions the existence of pure PM. Isolated weakness of neck flexors sparing finger flexors is consistent with PM. Furthermore, the favourable response to immunosuppression is again more typical of PM. Authorities have indeed claimed that the most distinguishing feature of IBM is resistance to immunosuppression [6]. Review of both patients’ biopsies (vastus lateralis) again confirmed the diagnoses, and specifically in Patient B’s biopsy, the absence of rimmed vacuoles. It is, however, acknowledged that changes of IBM may remain undetected as disease may be patchy.

Muscle histology has been correlated with clinical features and outcome in PM and IBM [7]. Review of 107 patients with initial histological diagnoses of PM and IBM revealed that 27 patients had PM (using clinical and pathological criteria), 64 had IBM and 16 of the remaining 43 (37%) had biopsy features of PM but clinical features of IBM, thus challenging the sensitivity of histological criteria for PM. Notably, Patient B, without classical clinical features of IBM, would also have been classified as PM.

The strongest evidence for genetic susceptibility to disease is linked with HLA alleles, and HLA B8 and DRB1*0301 are both associated with sporadic IBM in Caucasians [8]. These alleles are carried on the 8.1 ancestral haplotype (A1-B8-DR3-DQ2), a genetic risk factor for a many immune-mediated diseases [9]. TNF and C4, located in the MHC central region between HLA-B and DRB1, are important candidates as the 8.1 haplotype carries alleles associated with both high TNF and low C4 levels [9]. Furthermore, recombinant haplotype mapping has suggested that the primary HLA association with sporadic IBM lies in the MHC central region [10]. HLA typing for the monozygotic twins presented revealed A1,3, B8,51, DRB1*0101. Therefore, while DR3 was absent, the presence of the 8.1 central region cannot be excluded. Opportunities for recombinant haplotype mapping, as provided herein, may further our insight into genetic susceptibility to disease.

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A case of Yersinia enterocolitica mimicking Kawasaki disease

Sir, Kawasaki disease (KD) is the most common cause of acquired heart disease in children in the USA. Despite its description over four decades ago, the diagnosis remains a clinical one with many complex algorithms for atypical presentations [1]. Given its consequences when untreated, it is a serious diagnosis. Nevertheless, many infectious and inflammatory conditions are known to cause a clinical syndrome similar to KD. We describe here the first reported case of Yersinia enterocolitica mimicking KD in a 15-month-old girl.

A 15-month-old female presented with 10 days of irritability, fever up to 40.4°C and a diffuse erythematous rash. The parents also noted bright red lips and a suspicious lump in the side of her neck. On Day 2 of her illness, the family noted loose, non-bloody diarrhoea and fever up to 38.2°C. Her physical examination revealed an erythematous, maculopapular rash on her thigh, trunk, arms, palms, soles and, very minimally, on the face. Neurological examination was normal. Laboratory findings are summarized in Table 1. An ECG was within normal limits as was an echocardiogram.

By clinical and laboratory criteria, she fitted the diagnosis of incomplete KD and was admitted for intravenous immunoglobulin (IVIG) that she received on hospital Day 1. She remained febrile and required a second dose of IVIG on hospital Day 3. Fevers up to 39.9°C persisted at which point she received pulse dose steroids of methylprednisolone 30 mg/kg. After receiving one dose, she defervesced for a 24-h period. In the meantime, she had been started on high-dose aspirin for cardioprotection. Once afibrile, she was discharged on low-dose aspirin, prednisolone and famotidine. Of note, her persistent diarrhoea was thought to be part of the Kawasaki syndrome.

The patient presented one day after discharge with worsening diarrhoea and fever up to 38.2°C. Her physical examination was unchanged other than slightly increased irritability. Repeat laboratory findings are summarized in Table 1.

The working diagnosis continued to be KD and the patient was restarted on high-dose steroids. However, given the atypical presentation of an atypical disease, we pursued other diagnoses. A full rheumatological panel—ANCA, IgA, ANA and anti-β2 glycoproteins—was negative. Stool studies were repeated including Clostridium difficile toxin and a stool culture. Metronidazole was started for a presumptive diagnosis of C. difficile colitis. The day after the metronidazole was started, the patient defervesced. On hospital Day 11, her repeat stool cultures were positive for Y. enterocolitica: At this point, the metronidazole was discontinued and empiric treatment with sulphamethoxazole/trimethoprim was started. Her diarrhoea quickly subsided and she continued to be afibrile. She completed a 14-day course of treatment and did well.

**Table 1. Laboratory data**

<table>
<thead>
<tr>
<th>Test</th>
<th>Hospital Day 1</th>
<th>Hospital Day 11</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC (&gt;10000/µl)</td>
<td>15 (55% N, 31% L, 12% M)</td>
<td>30 (64% N, 7% B, 14% L)</td>
</tr>
<tr>
<td>Hgb (g/dl)</td>
<td>11.6±3.4</td>
<td>9.8±2.9</td>
</tr>
<tr>
<td>Platelet count/µl</td>
<td>502 000</td>
<td>597 000</td>
</tr>
<tr>
<td>CRP (mg/dl)</td>
<td>45</td>
<td></td>
</tr>
<tr>
<td>ESR (mm/hr)</td>
<td>86</td>
<td></td>
</tr>
<tr>
<td>Albumin (g/dl)</td>
<td>3.1</td>
<td>2.7</td>
</tr>
<tr>
<td>Bicarbonate (mg/dl)</td>
<td>19.2</td>
<td>18</td>
</tr>
<tr>
<td>Anion gap</td>
<td>14</td>
<td>13</td>
</tr>
<tr>
<td>DFA for adenovirus</td>
<td>Negative</td>
<td>Positive Y. enterocolitica</td>
</tr>
<tr>
<td>Rotavirus Ag</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>Clostridium difficile toxin</td>
<td>Negative</td>
<td>Negative</td>
</tr>
</tbody>
</table>

WBC: white blood cell count; N: neutrophils; L: lymphocytes; M: monocytes; B: bands; Hgb: haemoglobin; hct: haematocrit; DFA: direct fluorescence antibody test for viruses; Ag: antigen.