Relapsing Polychondritis and Malignant Lymphoma

Is Polychondritis Paraneoplastic?

Teruki Yanagi, MD; Tetsuri Matsumura, MD, PhD; Ryuta Kamekura, MD; Noriko Sasaki, MD; Satoshi Hashino, MD, PhD

Background: Relapsing polychondritis (RP) is associated with other rheumatic or autoimmune disease in about 30% of cases; however, an association with malignancy is rare with the exception of myelodysplastic syndrome (MDS).

Observation: Herein we report the first case, to our knowledge, of RP following splenic non-Hodgkin lymphoma (NHL), and we have reviewed all the previous well-documented reports that described the cases of RP associated with malignant lymphoma (ML).

Conclusions: Our case and the review of reported cases showed that RP preceded ML in 2 cases, RP occurred after diagnosis and treatment of ML in 2 cases, and RP and ML occurred simultaneously in 1 case. The types of ML encountered were Hodgkin lymphoma, orbital mucosa associated lymphoid tissue type lymphoma, nodal NHL, and splenic NHL. From the frequent association of RP with MDS and, less frequently, with ML, we speculate that some RP cases may occur as a paraneoplastic condition of the concurrent hematological malignancies.

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To date, there have been 4 case reports that have suggested an association between RP and lymphoma (Table).\(^5\)\(^6\) Our case and the review of reported cases showed that RP preceded lymphoma in 2 cases, RP occurred after diagnosis and treatment of lymphoma in 2 cases, and RP and lymphoma occurred simultaneously in 1 case. The types of lymphoma encountered were Hodgkin lymphoma, orbital mucosa-associated lymphoid tissue type lymphoma, nodal NHL, and splenic NHL. Yet, the association of RP with MDS has been described on several occasions, and it has been suggested that up to a quarter of RP cases may be associated with MDS.\(^2\) Only 5 cases in the world literature, including our case, cannot provide the evidence that there is a true association between RP and lymphoma. However, from the frequent association of RP with MDS and, less frequently with lymphoma, we speculate that some RP cases may occur as a paraneoplastic condition of the concurrent hematological malignancies, which was proposed by the case records of Massachusetts General Hospital.\(^7\)

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Correspondence: Teruki Yanagi, MD, Department of Dermatology, Tonan Hospital N1W6, Chuo-ku, Sapporo 060-0001, Japan (yanagi@med.hokudai.ac.jp).

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REFERENCES


Table. Clinical Features of Patients With Relapsing Polychondritis (RP) and Malignant Lymphoma (ML)

<table>
<thead>
<tr>
<th>Patient/Sex/Age at Onset of RP, y</th>
<th>Type of ML</th>
<th>ML Occurred Before/After RP</th>
<th>Manifestation of RP</th>
<th>Other Complication</th>
<th>Source</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/46</td>
<td>Hodgkin lymphoma Nodal NHL (diffuse, mixed cellular type)</td>
<td>6 mo after RP Simultaneously</td>
<td>Auricular swelling</td>
<td>Saddle nose deformity, thickened ears, deafness</td>
<td>. . . Miller et al(^5)</td>
</tr>
<tr>
<td>2/M/63</td>
<td>Hodgkin lymphoma stage IIIB</td>
<td>21 y before RP</td>
<td>Auricular swelling</td>
<td>Acute leukemia</td>
<td>Case records of MGH(^7)</td>
</tr>
<tr>
<td>3/M/41</td>
<td>Hodgkin lymphoma</td>
<td>12 y after RP</td>
<td>Auricular swelling, joint involvement</td>
<td>. . .</td>
<td>Lichauco et al(^8)</td>
</tr>
<tr>
<td>4/F/41</td>
<td>Orbital MALT type B cell lymphoma</td>
<td>3 y before RP</td>
<td>Auricular swelling, ocular, joint involvement</td>
<td>Nonspecific colitis</td>
<td>. . .</td>
</tr>
<tr>
<td>Present case/F/60</td>
<td>Extranodal NHL (splenic diffuse large B) stage IIA</td>
<td></td>
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</tr>
</tbody>
</table>

Abbreviations: MALT, mucosa-associated lymphoid tissue; MGH, Massachusetts General Hospital; NHL, non-Hodgkin lymphoma.