A case of juvenile myelomonocytic leukemia presenting with a B-lymphoblastic immunophenotype

Juvenile myelomonocytic leukemia (JMML) represents no more than 2% of leukemias in children. To facilitate the diagnosis of this pathology, centralized diagnostic facilities have been established in our country. The validity of such facilities was confirmed in a case of monosity associated with an immunophenotype suggestive of B-lymphoblastic leukemia.

The classification of myelodysplastic syndromes (MDS) in childhood has been the subject of controversy during the last decade. Although some investigators have argued that childhood MDS can be classified into three subgroups as the French-American-British (FAB) nomenclature for adult cases, others have pointed out that this is rarely used in practice.1 Children with MDS are subdivided into two groups, i.e. those with a more adult-type MDS and those suffering from a disorder with myeloproliferative features primarily observed in infancy and early childhood. The latter pathology is characterized by prominent hepatosplenomegaly, frequent skin involvement, leukocytosis, monosity, and the presence of immature precursors in peripheral blood and has traditionally been described as chronic myelomonocytic leukemia (CMML).2,3 or juvenile chronic myelogenous leukemia (JCML).4 The new term JMML has recently been proposed to avoid further confusion.5

A 15-month-old child with panniculitis of the right leg was referred to our Pediatric department. Peripheral blood analysis showed: Hb 9.5 g/dL, platelets 231 × 10^9/L, WBC 20.8 × 10^9/L, and the presence of monosity (25.4%; 5.2 × 10^10/L) with dysmorphologic features. Blood chemistry values were within normal ranges and viral serology was negative. The patient also had axillary lymphadenopathy and hepatosplenomegaly. Flow cytometric analysis showed the presence of an abnormally high percentage of B-lymphocytes (40% CD19+) in peripheral blood, 18% of which were positive for co-expression with CD10 antigen, frequently observed in common lymphoblastic acute leukemia. Fever ranging from 37°C to 38°C was also present and resolved by antibiotics. The suspicion of an oncohematologic disease indicated the need for a close follow-up and a bone marrow examination done after a month showed the presence of a high percentage of B-lymphocytes CD10+/CD19+, poor representation of hematopoietic series with slight dysmyelopoiesis and 2% of blasts. Spleenomegaly, leukocytosis (WBC 17.8 × 10^9/L) with monosity (24.1%) and circulating myeloid precursors were still present. Although other hematologic criteria of JMML were missing (bone marrow hypercellularity with less than 20% myeloid blasts and >10% of HbF),6,7 the probability of this diagnosis began to emerge. As the Italian National Registry, was demonstrated in this case of JMML presenting with an immunophenotype suggestive B-lymphoblastic leukemia.

<table>
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<tr>
<th>Patient BM MNC</th>
<th>175</th>
<th>130</th>
<th>49</th>
<th>31</th>
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</thead>
<tbody>
<tr>
<td>Normal BM MNC</td>
<td>35-190</td>
<td>0</td>
<td>30-95</td>
<td>0</td>
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<tr>
<td>Patient PB MNC</td>
<td>192</td>
<td>178</td>
<td>78</td>
<td>116</td>
</tr>
<tr>
<td>Normal PB MNC</td>
<td>18-178</td>
<td>0</td>
<td>20-77</td>
<td>0</td>
</tr>
</tbody>
</table>

Key words: juvenile myelomonocytic leukemia, JMML, B-lymphoblastic acute leukemia.

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