Letters to the Editor

Table 1. α-thal genotypes and associated hematologic values of 67 thalassemia patients.

<table>
<thead>
<tr>
<th>Subject</th>
<th>Mean Hb (g/dL)</th>
<th>Mean HbA2 (%)</th>
<th>Mean MCV (fL)</th>
<th>Mutation</th>
</tr>
</thead>
<tbody>
<tr>
<td>23</td>
<td>14.1±1.0</td>
<td>2.4±0.6</td>
<td>73.9±6.4</td>
<td>αβ2/αα</td>
</tr>
<tr>
<td>9</td>
<td>13.2±1.4</td>
<td>2.6±0.7</td>
<td>69.3±3.1</td>
<td>α-β/αα</td>
</tr>
<tr>
<td>3</td>
<td>13.2±1.6</td>
<td>3.1±1.4</td>
<td>68.3±4.8</td>
<td>αβ-β/αα</td>
</tr>
<tr>
<td>2</td>
<td>11.7±0.6</td>
<td>3.7±0.3</td>
<td>70.8±6.7</td>
<td>αβ2/αα</td>
</tr>
<tr>
<td>1</td>
<td>10.7</td>
<td>3.5</td>
<td>59.2</td>
<td>αβ2/ββ</td>
</tr>
<tr>
<td>1</td>
<td>13.6</td>
<td>3.8</td>
<td>66.0</td>
<td>αβ2/ββ</td>
</tr>
<tr>
<td>1</td>
<td>12.0</td>
<td>3.4</td>
<td>77.4</td>
<td>αβ2/αα</td>
</tr>
<tr>
<td>1</td>
<td>11.4</td>
<td>2.4</td>
<td>68.3</td>
<td>αβ-β/αα</td>
</tr>
<tr>
<td>1</td>
<td>not available</td>
<td>3.4</td>
<td>76.5</td>
<td>αβαα/αα</td>
</tr>
<tr>
<td>1</td>
<td>14.8</td>
<td>3.5</td>
<td>78.4</td>
<td>αβαα/αα</td>
</tr>
<tr>
<td>24</td>
<td>13.0±2.3</td>
<td>3.4±0.6</td>
<td>72.7±6.8</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Table 1. α-thal genotypes and associated hematologic values of 67 thalassemia patients.

α-globin gene deletion and point mutation analysis among Iranian patients with microcytic hypochromic anemia

We tested 67 Iranian individuals, presenting with low mean corpuscular volume (MCV) and mean corpuscular hemoglobin (MCH) levels, normal hemoglobin electrophoresis and iron status, for the presence of twelve common α-thalassemia gene deletions and point mutations. Five different mutations (−αβ2, −αβ2, −α2, −αβ2, Hb Constant Spring) were identified in a total of 43 cases.

References

Among the 67 Iranian subjects analyzed, α-globin gene mutations were identified in a total of 43 cases (64.2%). The most common α-thal genotypes were the heterozygous (α^+/-) and the homozygous (α^-/α^-) forms of the common single gene deletion α^+/-, which were observed in 23 (34.3%) and 9 (13.4%) subjects, respectively. Furthermore, 3 subjects (4.5%) were α^-/α^-/α^- (C. CSA), and 2 subjects (3.0%) were α^-/α^-/α^-/α^- (C. CSA). One individual each (1.5%) was found positive for the 4.2 deletion, 2.2% for the α^-/α^- deletion, 3.0% for the α^-/α^-/α^- deletion, 2.2% for α for Ha (constant Spring (α^-/α^-)), and 0.7% for the (-α^+) deletion.

α-thal is not prevalent as β-thal in Iran, but a significant number of cases with reduced MCV and MCH, normal Hb electrophoresis and normal iron status, are being referred to molecular biology clinics throughout the country as suspected cases of α-thal or normal Hb Aβ-thal. During earlier studies by our group, a high prevalence of the α^-/- single gene deletion among such patients was noted. In agreement with these preliminary data, our present study confirms that α^-/- is by far the most common cause of microcytic, hypochromic anemia in Iran. In addition, we observed other α-globin deletions and point mutations in considerably lower frequencies. We will now use DNS sequencing to investigate the patients without known mutations for possibly abnormal hemoglobins.

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References


Incidence and characteristics of myelodysplastic syndromes in Ourense (Spain) between 1994-1998

The very few reference epidemiological studies on myelodysplastic syndromes (MDS) have been carried out in Europe: Germany, France, UK and Sweden. We present the first Spanish study on the incidence and characteristics of MDS. The incidence rates, distribution by FAB subtypes, sex and age groups are given in the reference studies with minimal differences which we point out and attempt to explain.


The province of Ourense is located in the southeast of Galicia, Spain. It has 346,913 inhabitants, according to the 1996 Renovation Census. These inhabitants form a stable population with minimal migratory movements. There are no nuclear plants nor oil refineries in this region. The province’s hospital assistance is distributed between three centers: the Complexo Hospitalario de Ourense (CHOU), the Hospital Comarcal de Valdeorras and the Fundación Hospital Verin, the only provincial centers in which diagnosis and follow-up of MDS patients are carried out. The source for identifying the cases was the bone marrow databases at these hospitals. The sample consists of all the patients diagnosed with MDS according to the FAB classification criteria from January 1, 1994 up to December 31, 1998.

During the study period, 140 new cases of MDS were diagnosed. The crude incidence rate was 8.07/100,000/year (9.11 in men and 7.1 in women). The age standardized incidence rates were: 0 for age <50, 4.4 for age 50–59, 6.8 for age 60–69, 25.5 for age 70–79 and 56.7 for age >79. The incidence and the sex ratio increased with age; therefore, in the ≥70 year old age group, the incidence in men is 48.1/100,000/year, whereas in women it is 29.4/100,000/year (Table 1). The mean age at diagnosis was 77.5 years (CI 95%: 75.9, 79.0) with no significant differences between sexes (men 76.9 years and women 78.1 years, p=0.45) nor between FAB subtypes. Eighty-

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