Multiple myeloma in sickle cell syndromes

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Abstract—Multiple myeloma (MM) is rare among patients with sickle cell syndromes (SCS). We describe six Greek sickle cell patients aged 56 to 65 years: five haemoglobin Sβ+thalassaemia (HbSβ+thal), one sickle cell anaemia (HbSS), who developed MM (three IgGκ, one IgGλ, one IgAκ, and one IgGκ-IgAκ (biclonal). Our HbSβ+thal cases, represent the first reported association of this entity with MM. Generalized bleeding diathesis, stroke, grand mal seizures, bone marrow necrosis and other clinical manifestations due to hyperviscosity aggravated by sickle cell vasoocclusion were treated by plasmaphereses and exchange blood transfusions. The increase of mean survival in SCS patients due to the current medical facilities may have an impact on the incidence of MM among them, if a pathogenetic link between the two conditions exists. All our patients carried a diagnosis of cholelithiasis which may predispose to MM; two of them progressed from a monoclonal gammopathy of undetermined significance (MGUS) to MM. Further studies are needed in order to understand the relationship between SCS and MM.

Key words: Multiple myeloma; monoclonal gammopathy; sickle cell disease.

INTRODUCTION

Multiple myeloma (MM) is considered to be rare in association with sickle cell syndromes (SCS); only four cases are reported in the literature, namely, one black female from Jamaica [1], two black males from the USA [2, 3] and one male from India [4]. Analogous cases are not reported from the Mediterranean countries or from Africa where SCS have a high prevalence.

We describe six Greek SCS patients who developed MM. To the best of our knowledge these cases represent the first reported association of MM with haemoglobin Sβ+ thalassaemia (HβSβ+thal).

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CASE REPORTS

Case 1

A 56 year old woman with confirmed HbSβ+ thal and an IgAκ monoclonal gammopathy of undetermined significance (MGUS), diagnosed 12 years before, was admitted in June 1987, for weakness, bleeding diathesis, jaundice, blurred vision and hepatosplenomegaly. Laboratory investigation revealed an IgAκ MM (Table 1) and hyperviscosity. Serum relative viscosity (SRV) was 4.9 U (normal 1.4–1.8 U). The patient was treated by plasmaphereses, exchange transfusions and cyclophosphamide/prednisone in six monthly courses. Fifteen months later she was readmitted with a severe hyperviscosity syndrome (SRV 11.9 U) with bleeding diathesis, grand mal seizures and bone marrow necrosis. She improved dramatically in 4 weeks following plasmaphereses and blood transfusions but she died three months later in a crisis of seizures at home (December 1989).

Table 1.
Main laboratory data in 6 Greek patients with sickle cell syndromes (SCS) and multiple myeloma (MM)

<table>
<thead>
<tr>
<th>Cases</th>
<th>Age (g/l)</th>
<th>Hb</th>
<th>Hb electrophoresis pattern</th>
<th>Serum M-protein (g/l)</th>
<th>Serum viscosity (normal 1.4–1.8 U) %</th>
<th>Bone marrow plasma cell infiltration (%)</th>
<th>Amyloid deposits</th>
<th>Skeletal lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>56</td>
<td>72</td>
<td>10%A, 79%S, 6%F, 5%A2</td>
<td>IgAκ (48)</td>
<td>11.9</td>
<td></td>
<td>90</td>
<td>–</td>
</tr>
<tr>
<td>2</td>
<td>57</td>
<td>97</td>
<td>12%A, 76%S, 8%F, 4%A2</td>
<td>IgGκ (70)</td>
<td>4.3</td>
<td></td>
<td>60</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>55</td>
<td>120</td>
<td>6%A, 79%S, 12%F, 3%A2</td>
<td>IgGκ (65)</td>
<td>4.1</td>
<td></td>
<td>65</td>
<td>–</td>
</tr>
<tr>
<td>4</td>
<td>61</td>
<td>107</td>
<td>12%A, 68%S, 16%F, 4%A2</td>
<td>IgGκ (26)</td>
<td>3.2</td>
<td></td>
<td>50</td>
<td>–</td>
</tr>
<tr>
<td>5</td>
<td>42</td>
<td>75</td>
<td>88.2%S, 10%F, 1.8%A2</td>
<td>IgGκ (34)</td>
<td>not measured</td>
<td></td>
<td>35</td>
<td>–</td>
</tr>
<tr>
<td>6</td>
<td>65</td>
<td>56</td>
<td>20%A, 74%S, 1.6%F, 4.2%A2</td>
<td>IgGλ (26.6)</td>
<td>3.7</td>
<td></td>
<td>30</td>
<td>+</td>
</tr>
</tbody>
</table>