Epidemiology of Behçet’s disease using the Registry of Rare Diseases of Tuscany

INTRODUCTION

Behçet’s Disease (BD) is a systemic, chronic-relapsing vasculitis of unknown etiology whose diagnosis is based on clinical criteria. The BD clinical presentation is extremely variable, and its effective management requires the prompt initiation of immunosuppressive treatments to avoid irreversible damage to vital organs.

AIMS

To describe the epidemiology of BD in Tuscany (population: 3.8 million inhabitants) (Italy)

METHODS

BD cases collected by the population-based Registry of Rare Diseases were linked to registry of mortality, hospital discharge data, and medicine prescriptions data. Epidemiological indicators and causes of hospitalizations were assessed through a multi-source approach.

RESULTS

Registry of rare diseases of Tuscany (RTMR)

Hospital Discharge data

Registry of Mortality

Medicine prescriptions data

Comorbidity

Prevalence (2000-2014)

Complications as a function of therapy

In progress

<table>
<thead>
<tr>
<th>Causes of hospitalization (ICD9 CM codes)</th>
<th>TOT (n=140) (95%CI)</th>
<th>M (n=66) (95%CI)</th>
<th>F (n=74) (95%CI)</th>
<th>Diff proportion test (p value)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infectious And Parasitic Diseases (001-139)*</td>
<td>8 (6.9)</td>
<td>5 (9.6)</td>
<td>3 (4.7)</td>
<td>0.300</td>
</tr>
<tr>
<td>Neoplasms (140-239)</td>
<td>20 (17.2)</td>
<td>4 (7.7)</td>
<td>16 (25.0)</td>
<td>0.014</td>
</tr>
<tr>
<td>Endocrine, Nutritional And Metabolic Diseases, And Immunity Disorders (240-279)</td>
<td>37 (31.9)</td>
<td>11 (21.2)</td>
<td>26 (40.6)</td>
<td>0.026</td>
</tr>
<tr>
<td>Diseases Of The Blood And Blood-Forming Organs (280-289)</td>
<td>16 (13.8)</td>
<td>4 (7.7)</td>
<td>12 (18.8)</td>
<td>0.085</td>
</tr>
<tr>
<td>Diseases Of The Nervous System And Sense Organs (320-389)</td>
<td>46 (39.7)</td>
<td>17 (32.7)</td>
<td>29 (45.3)</td>
<td>0.168</td>
</tr>
<tr>
<td>Diseases Of The Nervous System (320-359)</td>
<td>27 (23.3)</td>
<td>6 (11.5)</td>
<td>21 (32.8)</td>
<td>0.007</td>
</tr>
<tr>
<td>Disorders Of The Eye And Adnexa (360-379)</td>
<td>22 (19.0)</td>
<td>12 (23.1)</td>
<td>10 (15.6)</td>
<td>0.306</td>
</tr>
<tr>
<td>Diseases Of The Circulatory System (390-459)</td>
<td>53 (45.7)</td>
<td>25 (48.1)</td>
<td>28 (43.8)</td>
<td>0.644</td>
</tr>
<tr>
<td>Diseases Of The Respiratory System (460-519)</td>
<td>27 (23.3)</td>
<td>10 (19.2)</td>
<td>17 (26.6)</td>
<td>0.348</td>
</tr>
<tr>
<td>Diseases Of The Digestive System (520-579)</td>
<td>46 (39.7)</td>
<td>19 (36.5)</td>
<td>27 (42.2)</td>
<td>0.533</td>
</tr>
<tr>
<td>Diseases Of The Genitourinary System (580-629)</td>
<td>38 (32.8)</td>
<td>14 (26.9)</td>
<td>24 (37.5)</td>
<td>0.226</td>
</tr>
<tr>
<td>Diseases Of The Skin And Subcutaneous Tissue (680-709)</td>
<td>20 (17.2)</td>
<td>8 (15.4)</td>
<td>12 (18.8)</td>
<td>0.630</td>
</tr>
<tr>
<td>Diseases Of The Musculoskeletal System And Connective Tissue (710-739) **</td>
<td>36 (31.0)</td>
<td>13 (25.0)</td>
<td>23 (35.9)</td>
<td>0.207</td>
</tr>
</tbody>
</table>

* excluded 1361 (Behcet's disease)
** excluded Orthopedic causes (traumatic or similar)

CONCLUSIONS

✓ Incidence and prevalence estimates are consistent with other European studies
✓ Age at diagnosis and at onset are higher in women (p<0.05)
✓ The study confirms the involvement of neurologic, vascular, and eye organs
✓ A higher percentage of hospitalizations for women was observed in all the causes except for diseases of the eye and the circulatory system

The use of a high-quality population-based rare disease registry, linkable to other health data sources, allows to provide a wide epidemiological profile of the Behçet’s disease.