THE PREVALENCE OF RHUPUS SYNDROME IN A MONOCENTRIC TERTIARY REFERRAL COHORT

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Background: Systemic lupus erythematosus (SLE) and rheumatoid arthritis (RA) are two systemic chronic autoimmune diseases characterised by a wide spectrum of clinical and immune-pathological features. In clinical practice, the concept of Overlap Syndrome is defined when the two diseases coexist in the same patient. Rhupus syndrome is a rare clinical condition, characterized by overlap of SLE and RA. However, it is still subject to debate whether the syndrome should be considered a distinctive entity or an aggressive form of SLE. To date, the estimated prevalence of rhupus is unknown.

Objectives: To evaluate the prevalence of patients with rhupus syndrome in our tertiary care center.

Methods: We chart-reviewed all patients with a diagnosis of SLE[1] or RA[2], based on the current ACR classification criteria, who presented to our outpatient clinic rheumatology unit. Clinical and laboratory data were retrospectively collected from patient notes. Diagnosis were systematically analyzed by two independent reviewers (SGF and ER). Disagreements were resolved by consensus; if consensus could not be achieved, a third party (SS) would provide an assessment of eligibility.

Results: One-hundred eighty-two patients with a diagnosis of SLE and 253 patients of RA, were identified. Out of 182 SLE and 253 RA cases analysed, only 2 patients fulfilled rhupus criteria. The estimated prevalence in our cohort was 0.46% (2/437).

In Figure 1 are illustrated the clinical manifestations of SLE (Panel A; malar rash) and RA (radiograph-documented erosive polyarthritis) of the two patients.

Additionally, we identified 7 patients (1.61% of the total cohort) that only partially fulfilled both criteria for SLE and RA. In detail, 1 patient (0.23%) manifested with an overlap of clinical manifestations of SLE and RA, while 1 patient had an overlap of both laboratory criteria. Two patients (0.46%) presented with clinical criteria of RA and laboratory criteria for SLE and RA and 3 patients (0.69%) with clinical criteria of SLE and laboratory criteria for SLE and RA.

Patient’s selection is resumed in Figure 2.

Figure 1

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Patient’s selection is resumed in Figure 2.

Patients’ fulfilling the diagnosis of rhupus is resumed in Table 1.

<table>
<thead>
<tr>
<th>Patient</th>
<th>ANA 1:320</th>
<th>RF (U/ml)</th>
<th>anti-dsDNA</th>
<th>anti-APCA (U/ml)</th>
<th>Articular involvement</th>
<th>SLE extra-articular manifestations</th>
<th>Immunosuppressive treatment undergone</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Homogenous pattern</td>
<td>219</td>
<td>positive</td>
<td>63</td>
<td>erosions of MTF and ulnar deviation of the wrists</td>
<td>leukopenia, malar rash</td>
<td>hydroxychloroquine, methotrexate, infliximab, sulfasalazine, ciclosporin, azathioprine, abatacept, rituximab, tocilizumab</td>
</tr>
<tr>
<td>2</td>
<td>Granular pattern</td>
<td>55</td>
<td>Positive</td>
<td>47.95</td>
<td>loss of ICs, CMCs and MCPs joint space and bilateral MCP joint erosions (2nd-4th)</td>
<td>Myelitis, leukopenia, malar rash, oral ulcers</td>
<td>hydroxychloroquine, methotrexate, azathioprine, mycophenolate, tocilizumab</td>
</tr>
</tbody>
</table>
Venous thrombosis involved 20 patients (90%). They were deep in 16 patients, superficial in one patient and superficial in 3 patients. It was recurrent in 14 patients. The most common site of deep vein thrombosis was the lower limbs (n=17). The others had unusual sites: inferior vena cava thrombosis (n = 3), superior vena cava thrombosis (n = 2), sub-hepatic vein (n=1) and upper limb thrombosis (n = 1). Pulmonary embolism was noted in one case.

Arterial damage involved 4 patients (20%) divided into arterial aneurysms (3 cases) and arterial thromboses (2 cases). The locations were divided as follows: lower extremity arteries in 2 cases, upper limb arteries in one case, pulmonary artery in 2 cases and abdominal aorta in one case. Lower limb ischemia had occurred in one patient.

Two patients (10%) had mixed vascular, arterial and venous involvement, confirming the multiple and ubiquitous nature of Beqehet’s disease.

The treatment was based on colchicine, anticoagulants and corticosteroids in venous thromboses. Immunosuppressive therapy was started in 4 patients in front of the unusual site.

In arterial cases, corticosteroids in combination with immunosuppressant were prescribed. Flattening of the aneurysm was indicated in 2 cases with simple operative follow-up.

Conclusion: Behçet disease (BD) is very common in the Mediterranean basin, mainly affects the 30-year-old man. This systemic disease is characterized by oral aphthosis, genital ulcers and systemic involvement including ocular, gastrointestinal, neurological, and vessels that make the severity of the disease.

All types of vessels, regardless of size and seat, may be affected, with venous tropism.

Our study has just supported current literature data regarding the extensive and recurrent venous thrombosis during MB. The seriousness of this venous involvement lies in the involvement of the cavae veins and in pulmonary embolism.

Arterial damage, such as thrombosis and/or aneurysm, is rare but maybe life-threatening.

Early diagnosis, intensive and appropriate treatment, regular follow-up and the involvement of a multidisciplinary team including internists, vascular surgeons and radiologists are key to better management of patients with angiobehçet.

Our study illustrates the frequency and significance of vascular involvement in BD.

Disclosure of Interests: None declared