POTENTIAL PREDICTORS OF VISCERAL INVOLVEMENT IN ADULT IGA VASCULITIS

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Background: Predictors of severity of visceral involvement in acute adult IgA vasculitis (IgAV) are poorly recognised.

Objectives: The aim of our study was to evaluate the role of smoking and extension of skin lesions on the visceral manifestations of acute adult IgA vasculitis.

Methods: We analysed medical records of adult, histologically proven IgAV cases, diagnosed at our secondary/tertiary rheumatology centre between 1 January 2010 and 31 December 2017. Purpura was defined as generalised when skin lesions extended above the waistline. Gastrointestinal (GI) disease was considered severe in case of bloody diarrhoea, ileus or bowel perforation. Renal disease was defined as severe when nephritic syndrome with acute renal failure or nephrotic syndrome developed.

Results: During the study period we identified 230 incident IgAV cases (57.8% males, median (IQR) age 64.8 (45.6–77.3) years). Ninety-eight (42.6%) patients were smokers (56 past and 42 current). Skin, joint, GI, renal and involvement were present in 230 (generalised purpura in 114 (49.6%), necrotising in 108 (47.0%), 93 (40.4%), 70 (30.4%); severe in 17 and 102 (44.3%; severe in 27) patients, respectively. Smoking was associated with renal disease (RR 1.3 (95% CI 1.0–1.8)) and its severity (RR 3.2 (95%CI 1.5–7.0)), but not with GI involvement or its severity. Generalised purpura was associated with GI involvement (RR 2.9 (95%CI 1.8–4.7) and its severity (RR 3.3 (95%CI 1.1–9.8)), as well as with renal involvement (RR 1.4 (95% CI 1.0–1.9)). Data of combined influence of smoking and purpura extension on visceral involvement are presented in table 1. The risk of severe renal involvement in IgAV was the highest in ever-smoker with generalised purpura (RR 8.1 (95%CI 1.9–34.7) in comparison to IGAV non-smoker with localised purpura).

Table 1 The influence of smoking and purpura extension on visceral involvement in IgAV

<table>
<thead>
<tr>
<th>Visceral involvement</th>
<th>Non-smokers with localised purpura</th>
<th>Ever-smokers with localised purpura</th>
<th>Non-smokers with generalised purpura</th>
<th>Ever-smokers with generalised purpura</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>69</td>
<td>47</td>
<td>63</td>
<td>51</td>
</tr>
<tr>
<td>GI (%)</td>
<td>13.0</td>
<td>19.1</td>
<td>47.6</td>
<td>43.1</td>
</tr>
<tr>
<td>Severe GI (%)</td>
<td>2.9</td>
<td>4.3</td>
<td>11.1</td>
<td>11.8</td>
</tr>
<tr>
<td>Renal (%)</td>
<td>30.4</td>
<td>46.8</td>
<td>47.6</td>
<td>56.9</td>
</tr>
<tr>
<td>Severe renal (%)</td>
<td>2.9</td>
<td>14.9</td>
<td>9.5</td>
<td>23.5</td>
</tr>
</tbody>
</table>

Legend: GI gastrointestinal; severe GI involvement: bloody diarrhoea or ileus or bowel perforation; severe renal involvement: nephritic syndrome with acute renal failure or nephrotic syndrome;

Conclusions: Smoking and generalised purpura were associated with visceral involvement in adult IgAV.

Disclosure of Interest: None declared


INSUFFICIENT IMMUNOSUPPRESSIVE USE IS THE LEADING CAUSE OF VASCULAR RELAPSES IN BEHÇET’S DISEASE

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Background: Vascular involvement is observed in up to 40% in Behçet Disease (BD) patients, as an important cause of mortality and morbidity, especially for males (Kural-Syahi E et al, 1984).

Objectives: Purpose of this study is to describe clinical-demographic properties, treatments and prognosis of vascular BD patients in a tertiary rheumatology clinic.

Methods: BD patients fulfilling ISG (1990) criteria are recruited from the multi-disciplinary Behçet’s Clinic in Marmara University, Istanbul for this retrospective study. All data is collected from patient files (ISG for BD, 1990).

Results: Mean age of BD patients (MF:102/22) was 29.3±7.3 years at diagnosis and 32.4±5.5 years during first vascular event. Median follow up was 47.7±71.8 months. Mean age of female patients was significantly older during first vascular event (table 1). 73.2% of vascular involvement was venous, mostly deep vein thrombosis (table 2). 32% (n=40) of patients presented first with a vascular event and diagnosed as BD. Twenty (16%) patients were diagnosed with a median of 121–120 months after the first vascular event. 15 (6.5%) patients were using immunosuppressive (IS- mainly azathioprin) drugs either for resistant mucocutaneous symptoms or major other organ involvement during the first vascular event. Vascular relapse rate was 40.7% and it was similar between sexes (F: 33.3% vs M: 42.2%, p=0.6). After the first vascular event, 96 (85.7%) patients had been treated with ISs and 58.9% used anticoagulants. Median IS and anticoagulant usage duration was 25.5 (5–48) and 2 (0–12) months respectively. Relaps rate was higher in patients who had stopped ISs (87.5% vs 32.3%). IS treatment duration

Disclosure of Interest: None declared

Our results show that female BD patients have a vascular event at a later age compared to males, but the course of vascular disease is not influenced with gender. Early termination of immunosuppressive treatments seems to be the most important cause of vascular relapses.

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Disclosure of Interest: None declared

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Methods: Retrospective national multicenter open-label study on 19 BD patients treated with apremilast at standard dose of 30 mg twice daily. The main outcome was achievement of oral ulcers remission.

Results: We included 19 patients (14 women and 5 men) with a mean age of 43.6±14.8 years. Before apremilast, all patients had also received several systemic conventional drugs: oral corticosteroids (n=18), colchicine (n=19), NSAIDs (n=10), methotrexate (n=10), azathioprine (n=10), cyclosporine (n=6), infliximab (n=3), adalimumab (n=5), dapsone (n=3), etanercept (n=1), mycophenolate mofetil (n=1), tocilizumab (n=1). The main clinical symptoms for starting apremilast were oral aphthous ulcers (n=19) and genital ulcers (n=14). Other manifestations present at apremilast onset were arthralgia/arthritis (n=6), folliculitis/pseudofolliculitis (n=6), asthenia (n=5), furunculosis (n=1), erythema nodosum (n=1), erythematous and scaly skin lesions (n=1), psoriasis (n=1), deep venous thrombosis (n=2) and ileitis (n=1). Table 1 shows the evolution of the patients. After a median follow-up of 6 (interquartile range, 5–10) months, most of the patients experienced clinical improvement. In this period of time, 11 patients developed any side-effect: dyspepsia (n=5), nausea (n=4), diarrhoea (n=4), abdominal pain (n=4), headache (n=3), loss of appetite (n=3), weight loss (n=1) and halo sickness (n=1). Three patients had to reduce the dose to 30 mg/day. Apremilast was discontinued in 4 patients: because of not obtaining the expected improvement (n=2), due to desire of pregnancy (n=1) and due to development of neurological involvement (n=1).

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