Objectives: Summarizing the clinical characteristics of IgG4-related diseases to improve understanding of clinicians and achieve early diagnosis and reasonable treatment.

Methods: The clinical data of 15 patients with IgG4-related diseases diagnosed from 2014 to 2016 were retrospectively analyzed.

Results: The ratio of male to female was 2:1 in 15 patients. The average age of onset was 56.8 years (34 to 84 years of age). From the first symptom to diagnosis of 8 days to 4.75 years, the most common first symptom in 15 patients was Submandibular gland swelling, pain, anorexia, cough, the most common affected organs were the pancreas, lungs, submandibular glands. There were 11 cases of histopathological examination by surgery or needle biopsy. The histopathology showed interstitial fibrous tissue hyperplasia with more lymphocyte and plasma cell infiltration. Immunohistochemistry showed that the number of IgG4-positive cells was 1:2:pHPPF, and IgG4/IgG was >40%. Two of the 15 patients underwent surgical resection of the affected organ lesions before definitive diagnosis. One patient was treated with mycophenolate mofetil and one patient was given a medium dose of hormone combined with tacrolimus. The remaining 13 patients were treated with simple hormones or combination immunosuppressive agents, including hydroxychloroquine, mycophenolate mofetil, azathioprine, and cyclophosphamide.

Conclusion: The organs involved in IgG4-related diseases are diverse, the clinical manifestations are not specific, and the hormone combined immunosuppressive therapy is effective.

REFERENCES


Disclosure of Interests: None declared


Abstract AB0629 Table 2. Comparison of CEC, CEP, aCEC and rCECs between groups

<table>
<thead>
<tr>
<th>Group</th>
<th>CEC</th>
<th>CEP</th>
<th>aCEC</th>
<th>rCEC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1</td>
<td>5.09 ± 4.7</td>
<td>13.49 ± 6.8</td>
<td>4.39 ± 5.78</td>
<td>6.44 ± 5.4</td>
</tr>
<tr>
<td>Group 2</td>
<td>2.52 ± 2.65</td>
<td>7.62 ± 5.6</td>
<td>2.24 ± 2.55</td>
<td>5.43 ± 3.49</td>
</tr>
<tr>
<td>Group 3</td>
<td>4.89 ± 3.33</td>
<td>15.6 ± 11.21</td>
<td>8.17 ± 10.1</td>
<td>9.03 ± 7.73</td>
</tr>
<tr>
<td>Group 4</td>
<td>4.09 ± 4.9</td>
<td>13.71 ± 0.042</td>
<td>8.78 ± 2.54</td>
<td>3.52 ± 2.34</td>
</tr>
</tbody>
</table>

Conclusion: Increased levels of aCECs may be an indicator of active vascular involvement in BD. But in the current study aCEC levels did not show a difference between groups but none of the patients had active vascular involvement which may cause of this sameness. CEP and resting CEC levels were elevated in both groups of patients with thrombosis. So CEC may be a marker for vascular damage but it is not specific for BD.

REFERENCES


Disclosure of Interests: None declared


Abstract AB0630 Table 1. Demographic features of the study population

<table>
<thead>
<tr>
<th>Group</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Group 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age mean ± standard deviation</td>
<td>43.55 ± 8.31</td>
<td>46.65 ± 9.43</td>
<td>42.50 ± 15.22</td>
<td>41.65 ± 6.54</td>
</tr>
<tr>
<td>Duration of disease</td>
<td>14.90 ± 8.69</td>
<td>11.30 ± 8.80</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>Male</td>
<td>Female</td>
<td>Male</td>
</tr>
<tr>
<td>n (%)</td>
<td>n (%)</td>
<td>n (%)</td>
<td>n (%)</td>
<td></td>
</tr>
<tr>
<td>Group 1</td>
<td>9 (% 45.0)</td>
<td>12 (% 60.0)</td>
<td>11 (% 55.0)</td>
<td>8 (% 40.0)</td>
</tr>
<tr>
<td>Group 2</td>
<td>11 (% 55.0)</td>
<td>14 (% 70.0)</td>
<td>14 (% 70.0)</td>
<td>6 (% 30.0)</td>
</tr>
</tbody>
</table>

Disclosure of Interests: None declared

of nodular panniculitis (including 14 cases of mesenteric involvement) and 3 cases of special type of panniculitis were collected. Patients with nodular panniculitis were treated with multiple subcutaneous nodules or masses. According to the affected parts, they are divided into skin type and system type. Among them, 10 cases are skin type and 42 cases are system type. The most common parts is mesenteric and kidney involvement. 9 of 55 patients was combined with tumor. Patients with mesenteric panniculitis mainly present abdominal pain and bisating, which are mainly diagnosed according to CT. Patients with systemic panniculitis can be significantly relieved after treatment with hormones and immunosuppressive agents.

Conclusion: Panniculitis is an inflammatory disease of unknown etiology. It is common in middle-aged and elderly people. It has various clinical manifestations and lacks specificity. The diagnosis is mainly based on pathological results. It is easy to be combined with tumors. When subcutaneous nodules are found, pathological examination should be improved in time in order to avoid misdiagnosis.

REFERENCES

Disclosure of Interests: None declared

AB0631

INTERSTITIAL LUNG DISEASE – A RARE MANIFESTATION OF MICROSCOPIC POLYANGIITIS
Ekaterina Vinogradova1,2, Nikolai Bulanov3, Elena Shchegoleva2, Larisa Akukina1, Anastasia Zvykovy1,2, Pavel Novikov2, Sergey Mosiev1,2, Lomonosov Moscow State University, Moscow, Russian Federation; 2Sechenov University, Moscow, Russian Federation

Background: Intestinal lung diseases (ILD) are a group of diffuse inflammatory and/or fibrotic lung disorders with similar clinical, radiologic and histopathologic features. The coexistence of ILD and ANCA-associated vasculitis has been reported in case reports and small case series.

Objectives: The aim of this study was to assess the prevalence, clinical and radiological characteristics of ILD in MPA patients admitted to the Rheumatology Department of our hospital.

Methods: This retrospective single center cohort study included 102 patients diagnosed with MPA according to CHCC 2012 and EMA algorithm. Rheumatology Department of our hospital.

Results: ILD-features on HRCT was found in 11 (11.8%) of 102 patients with MPA. Their median age was 55 [53; 63] years. All of them were ANCA-positive (Table 1). In 5 cases interstitial pneumonia was the first and the sole manifestation that preceded for the occurrence of overt systemic vasculitis for a median of 3 years. The radiologic patterns included non-specific interstitial pneumonia in 5 cases, usual interstitial pneumonia in 3 cases and unclassifiable interstitial pneumonia in 3 cases. The most common clinical manifestations were non-productive cough (82%), progressive dyspnea (82%) and crepitation (27%). The most common extrathoracic manifestations of MPA in patients with ILD were glomerulonephritis with decreased renal function (91%), fever (100%) and arthritis (91%). All patients received induction therapy with glucocorticosteroids combined with cyclophosphamide, rituximab, azathioprine or methotrexate.

Conclusion: ILD is a rare manifestation of MPA, which can precede systemic manifestations. ANCA-associated vasculitis should be included in the spectrum of differential diagnosis in patients with ILD.

Disclosure of Interests: None declared

AB0632

MAINTENANCE RITUXIMAB IN ANCA ASSOCIATED VASCULITIS – OBSERVATIONAL DATA FROM UNIVERSITY HOSPITAL COVENTRY, UK
Megan Rutter1, Shirish Dubey1, Andrew Short2. 1University Hospital Coventry and Warwickshire, Department of Rheumatology, Coventry, United Kingdom; 2University Hospital Coventry and Warwickshire, Department of Nephrology, Coventry, United Kingdom

Background: Rituximab is considered an effective maintenance therapy in anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV), with reduced relapse rates post induction therapy and comparable adverse event profile to other agents such as Azathioprine.

Objectives: To describe the clinical outcomes of all patients receiving maintenance Rituximab for AAV in University Hospital Coventry in UK.

Methods: Records from the Renal and Rheumatology units were used to identify patients receiving maintenance Rituximab. Electronic patient records (EPR) were used to collate laboratory and clinical data.