of nodular panniculitis (including 14 cases of mesenteric involvement) and 3 cases of special type of panniculitis were collected. Patients with nodular panniculitis often presented 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 bloating, 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 POLYANGITIS

Ekaterina Vinogradova1,2, Nikolai Bulanov3, Elena Shchegoleva4, Larisa Akulkina1, Anastasiia Zykova1,2, Pavel Novikov3, Sergey Mosieiev2, Lomonosov Moscow State University, Moscow, Russian Federation; 2Sechenov University, Moscow, Russian Federation

Background: Interstitial lung diseases (ILD) are a group of diffuse inflammatory and/or fibrotic lung disorders with similar clinical, radiological and histopathological 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. We assessed Birmingham Vasculitis Activity Score (BVAS) at disease onset and VDI (Vasculitis Damage Index) at the end of follow up in each patient. ANCA type and titer were assessed by enzyme-linked immunosorbent assay (ELISA). The results of repeated high resolution computed tomography (HRCT) of the chest were analyzed and interstitial changes (local ground-glass opacity, reticular changes, honeycombing, traction bronchiectasis) were classified in four patterns: non-specific interstitial pneumonia (NSIP), typical interstitial pneumonia, possible interstitial pneumonia, and "inconsistent" usual interstitial pneumonia (UIP).

Results: ILD-features on HRCT was found in 11 (11.8%) of 102 patients with MPA. Their median age was 55 [S3; 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 (91%), fever (100%) and arthritis (91%). All patients received induction therapy with glucocorticosteroids and immunosuppressive agents. The adverse event profile to other agents such as Azathioprine 2. Among them, 10 cases are skin type and 42 cases are system type.

Abstract AB0631 Table 1. Characteristics of patients with MPA and ILD

<table>
<thead>
<tr>
<th>n</th>
<th>11</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male gender, n (%)</td>
<td>7 (64)</td>
</tr>
<tr>
<td>Age of the onset of ILD, Me, IQR, years</td>
<td>55 [53; 63]</td>
</tr>
<tr>
<td>Age of the onset of MPA, Me, IQR, years</td>
<td>59 [54; 65]</td>
</tr>
</tbody>
</table>

ILD onset

Before systemic manifestations of MPA, n (%) 5 (45)
At MPA onset, n (%) 6 (55)

ANCA specificity

MPO-ANCA, n (%) 6 (55)
PR3-ANCA, n (%) 2 (18)
undetermined ANCA, n (%) 3 (27)

Clinical symptoms

Fatigue, n (%) 11 (100)
Fever, n (%) 11 (100)
Weight loss, n (%) 4 (36)
Arthralgia, n (%) 10 (91)
Skin vasculitis, n (%) 5 (45)
Renal manifestations, n (%) 10 (91)
Ear, nose and throat involvement, n (%) 4 (36)
Chronic cough, n (%) 9 (82)
Hemoptysis, n (%) 2 (18)
Dyspnea, n (%) 9 (82)
Crackles, n (%) 3 (27)

Laboratory and instrumental findings

Increasing of ESR and/or CRP, n (%) 10 (91)
Hemoglobin, g/dl 7 (64)
24-hours proteinuria, g/day 0.61 [0.25; 1.95]
Estimated GFR, ml/min/1.73 m² 39 [19; 60]

Radiological patterns

UIP, n (%) 3 (27)
NSIP, n (%) 5 (45)
Unclassified IP, n (%) 3 (27)
Mediastinal lymphadenopathy, n (%) 3 (27)
Emphysema, n (%) 2 (18)

Remission induction therapy

Glucocorticosteroids, n (%) 11 (100)
Cyclophosphamide, n (%) 7 (64)
 Rituximab, n (%) 1 (9)
 Azathioprine, n (%) 1 (9)
 Methotrexate, n (%) 1 (9)

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

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Background: Rituximab is considered an effective 1 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 2.

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.

<table>
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<td>Age of the onset of MPA, Me, IQR, years</td>
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</table>
Results: We identified 38 patients, with the diagnosis of AAV made 11 months to 19 years previously, 16 (42%) had been diagnosed within the last five years. Age range was 19-87 years old with roughly half (21; 54%) being male. Maintenance Rituximab was started between January 2010 and September 2018. Most patients received a fixed 6 monthly protocol. Seven patients (18%) died within the observation period. More details in table 1 below.

Table 1. Results

<table>
<thead>
<tr>
<th>Total number (n) patients</th>
<th>n (%)</th>
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<tbody>
<tr>
<td>Serology</td>
<td>n (%)</td>
</tr>
<tr>
<td>ANCA positive</td>
<td>21 (55.3%)</td>
</tr>
<tr>
<td>PR3 positive</td>
<td>9 (23.7%)</td>
</tr>
<tr>
<td>MPO positive</td>
<td>11 (28.9%)</td>
</tr>
<tr>
<td>Previous cyclophosphamide</td>
<td>20/36 (55.6%)</td>
</tr>
<tr>
<td>Indication for starting Rituximab</td>
<td>9 (23.7%)</td>
</tr>
<tr>
<td>Failure of cyclophosphamide</td>
<td>20 (52.6%)</td>
</tr>
<tr>
<td>Flare/worsening disease despite disease-modifying therapy</td>
<td>3 (7.9%)</td>
</tr>
<tr>
<td>Intolerant of other disease-modifying therapies</td>
<td>3 (7.9%)</td>
</tr>
<tr>
<td>Young patient of child-bearing potential</td>
<td>2 (5.3%)</td>
</tr>
<tr>
<td>Patient preference</td>
<td>1 (2.6%)</td>
</tr>
<tr>
<td>Not documented</td>
<td>1 (2.6%)</td>
</tr>
</tbody>
</table>

Concurrent medication

- 12 (31.6%)
- Azathioprine 11 (28.9%)
- Mycophenolate mofetil 7 (18.4%)
- Methotrexate 1 (2.6%)
- Hydroxychloroquine 1 (2.6%)
- Tacrolimus 12 (31.6%)
- Prednisolone monotherapy 1 (2.6%)
- Nil

IgM

- 9 (23.7%)
- Low IgG and IgM 17 (44.7%)
- Isolated low IgM 3 (7.9%)
- Not tested 2 (5.3%)
- Clinical response to Rituximab 31 (81.6%)
- Full 2 (5.3%)
- Partial: requiring addition of further disease-modifying therapy 2 (5.3%)
- Mild disease progression 3 (7.9%)
- Flare within first 2 years of treatment
- First 2 years of Rituximab treatment 11 (28.9%)
- Ongoing 24 (63.2%)
- Completed 3 (7.9%)
- Not completed 1 (2.6%)
- Discontinued due to infections 2 (5.3%)
- Patient died
- Subsequent treatment† (n=24) 6/24 (25.0%)
- Rituximab continued as primary therapy 6/24 (25.0%)
- Regular Rituximab restarted following flare 1/24 (4.2%)
- Mycophenolate mofetil 4/24 (16.7%)
- Azathioprine 2 (8.3%)
- Prednisolone monotherapy

Patient died 2/24 (8.3%) 5/24 (13.3%)

† Renal transplant patient
† In 2/3 (67%) patients associated with poor compliance/missed doses
† In patients who completed 2 years maintenance Rituximab

Conclusion: The effectiveness of Rituximab as a maintenance therapy in AAV is borne out by this real world data, with 31/38 patients demonstrating full clinical response. Longer term treatment and low grade disease continue to be issues as evidence for optimal management strategies is lacking. Infections, including severe infections necessitating admission, were common. PR3 positive patients are over-represented, in keeping with the known higher rate of relapse.

REFERENCES


Yes

Disclosure of Interests: Megan Rutter: None declared, Shirish Dubey Grant/research support from: Roche provided a grant 5 years ago, Speakers bureau: Yes, SMS and Internis, Andrew Short: None declared


AB0632B  
TB OR NOT TB, THAT IS THE VASCULITIC QUESTION

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Background: It is important to remember that there are many disorders that can mimic the clinical, radiographic and histological features of vasculitis. Tuberculosis (TB) is often referred to as the great imitator and therefore, needs to be considered early in the differential diagnosis of vasculitis.

Objectives: To emphasize the importance of being aware of noninflammatory mimics of vasculitis to be able to avoid unnecessary and potentially harmful immunosuppression.

Methods: We report a challenging case of a patient presenting with an inflammatory subglottic lesion.

Results: A 81-year-old lady initially presented to otorhinolaryngology (ENT) with hoarseness and and restriction of breathing. Nasendoscopy revealed a subglottic lesion with oedema and inflammation. Biopsy showed florid necrotising granulomatous inflammation. Upon questioning, patient revealed a one year history of epistaxis and nasal crusting requiring regular nasal douching. She also reported lack of energy, loss of appetite, but denied any rash, weight loss or night sweats. She suffered with a severely dry mouth that was causing difficulty swallowing. She also complained of an occasional cough with small amount of phlegm which she attribute to the difficulty with swallowing. Rheumatology team felt that this could be a possible localised Granulomatosis with polyangiitis and requested ANCA and a vasculitis screen.

Interestingly, the patient’s chest x-ray showed apical scarring, consistent with healed mycobacterial disease. A CT neck/thorax and abdomen was done which showed diffuse mucosal thickening of the subglottis and reticulonodular changes in the lungs. She was also noted to have deranged liver function test around this time. Gastroenterology advised that there was no clear cause for the transaminitis and an autoimmune screen was requested. Given the chest findings on CT, a sputum sample was sent for microscopy and culture. The patient had been started on dexamethasone on admission by ENT and reported substantial improvement in the hoarseness of voice and shortness of breath. ANCA then came back negative, as did the connective tissue screen. While the rheumatology team was considering how to further manage the possible vasculitis, the sputum smear came back positive for acid fast bacilli and the diagnosis was clinched: our patient was suffering with the extremely rare laryngeal tuberculosis, along with pulmonary TB. She was started on anti-tuberculous medication that have been introduced incrementally given the deranged liver function tests. The patient was doing well on her most recent follow up in the respiratory department.

Conclusion: Greater awareness of vasculitis mimics like TB and a high index of suspicion are needed when assessing a patient presenting with the protean manifestations of suspected vasculitis. It is therefore prudent to include TB in the differentials, whenever consider vasculitis.

REFERENCES


