Other orphan diseases

AB1073 A RETROSPECTIVE COHORT STUDY OF IGG-4 RELATED DISEASE IN IRIAN PATIENTS
Aeidi Al Ghaith1,2, Aurelie Fabre3, Eamonn S. Molloy2. 1Al Nahda Hospital, Department of Medicine/Rheumatology, Muscat, Oman; 2St. Vincent’s University Hospital, Rheumatology, Dublin, Ireland

Background: Immunoglobulin (Ig) G4-related disease (IgG4RD) is a novel clinical entity characterized by elevated serum IgG4 concentration and tumefaction or tissue infiltration by IgG4-positive plasma cells.

Objectives: To describe the clinical presentations, laboratory features, imaging manifestations, histopathologic characteristics and treatments in a cohort of 38 patients with IgG4RD.

Methods: A retrospective study was performed at St. Vincent’s University Hospital. Clinical, laboratory, imaging and histopathologic data was retrieved from electronic records. All data were assessed using SPSS 24.0.

Results: Median age was 59 years with M:F ratio= 2.2:1. 24 (63.2%) patients were between 25-65 years, 14 (36.8%) were >65 years. 23 (60.5%) patients fulfilled the Comprehensive Diagnostic Criteria for IgG4RD as ‘definite’, whereas 5 (13.2%) patients fulfilled ‘probable’ diagnosis and 10 (26.3%) patients fall in ‘possible’ category. GI manifestations (followed by pancreatic) were the most frequent clinical presentation. 23 (60.5%) patients presented with single organ involvement; pancreas was the most frequently involved organ (17/38, (44.7%)), 55.3% had a serum IgG4 level above 135mg/dL. Lymphoplasmacytic infiltration was the commonest histopathologic pattern reported in 29 (76.3%) specimens. 25 (65.8%) patients had received steroid therapy and 19 (50.0%) had a good response. 11 (28.9%) patients received immuno-modulatory agents including Rituximab (n=4), Azathioprine (n=7), and Mycophenolate mofetil (n=4). Overall, 28 (73.7%) patients had complete remission with treatment.

Conclusion: IgG4RD is a rare entity in Ireland and an inadequately understood condition overall. Further research is required to better understand the pathophysiology, clinical course and optimal treatment for IgG4RD.

REFERENCES


AB1074 AUTOIMMUNE/INFLAMMATORY SYNDROME INDUCED BY ADJUVANTS—ASIA—RELATED TO BIOMATERIALS: ANALYSIS OF 50 CASES
Jaume Alliotas-Reig1, Enrique Esteve-Valverde2, 1Systemic Autoimmune Disease Unit, Department of Internal Medicine, Vall d’Hebron University Hospital, Department of Medicine, Universitat Autonoma Barcelona,., Barcelona, Spain; 2Internal Medicine Department, Althaia Healthcare University Network of Manresa, Systemic Autoimmune Disease Unit, Manresa, Barcelona, Spain

Background: Systemic autoimmune or granulomatous disorders related to biomaterials of human use have rarely been described

Objectives: The aim of this study was to report cases of autoimmune/inflammatory syndrome induced by adjuvants (ASIA) related to biomaterial injections and prostheses, mainly silicone, hyaluronic acid, acrylamides and methacrylate compounds in a Spanish patient cohort.

Methods: This study is a retrospective analysis of clinical, laboratory, histopathological and follow-up data of 50 cases of patients suffering from late-onset, non-infectious autoimmune/autoimmune disorders related to biomaterials. Late onset was defined as 3 months or more post injection. Data were obtained through a further non-systematic but comprehensive review of the literature. Fifty cases of late-onset adverse reactions related to biomaterial injections or prostheses were reviewed.

Results: All cases had systemic complaints that could be categorised as ASIA. In all but five patients, inflammatory features at the implantation location were present. Forty-one patients (# 1–40; 82%) were male, and the age range was 21–84 years. The mean age was 52.1 ± 15 years. The mean duration of symptoms was 6 ± 9 months.

Conclusion: This study of ASIA and related to biomaterials is the first of its kind in Spain. Further prospective studies are needed for a more thorough analysis of biomaterial-related ASIA.
site preceded distant or systemic manifestations. Abnormal blood tests were common. Localised inflammatory nodules and panniculitis in 88.88% of patients, idiopathic orbital inflammatory syndrome, Sjögren’s syndrome, sarcoidosis, human adjuvant disease, vasculitis, inflammatory bowel syndrome and inflammatory polyradiculopathy. 11.11% cases presented primarily with systemic autoimmune disorders. Conclusion: Biomaterials and protheses can provoke late-onset systemic autoimmune disorders fulfilling ASIA criteria, or present primarily local/ regional inflammatory reactions that may eventually evolve into systemic autoimmune and/or granulomatous disorders which fall under ASIA.

REFERENCES

AB1075
IDIOPATHIC ORBITAL PSEUDOTUMOUR: A CASE SERIES AND LITERATURE REVIEW
Edna Amaya Cabrera, Sergio Corpa-Cruz, Veronica Gonzalez-Diaz, Gloria Martinez-Bonilla, Sergio Gutierrez-Ureña, Marisol Inguez Soto. Hospital Civil Fray Antonio Alcalde, Reumatología, Mexico, Mexico

Background: Idiopathic orbital pseudotumor (IOP), also known as idiopathic orbital inflammatory syndrome is a benign, non-infective, inflammatory condition of the orbit without identifiable local or systemic causes. After Grave’s disease and lymphoproliferative disorders, orbital pseudotumor is the 3rd most common ophthalmologic disease of the orbit and account for approximately 8-11% of all the orbital tumors. Pathogenesis of orbital pseudotumor remains elusive but several lines of evidence point to immunoadaptive processes as the likely underlying ocular mechanism. The etiology of orbital pseudotumor is unknown, but infection, autoimmune disorder, and aberrant wound healings have been put forward as possibilities. The ocular manifestations of orbital pseudotumor may include periorbital edema, erythema, proptosis, ptosis, diplopia and pain with eye movements.

Objectives: Describe clinical and demographic characteristics, most frequent diagnoses, immunological serology and treatments in patients with Orbital Pseudotumor.

Methods: We performed a retrospective cohort study of adult and pediatric patients with orbital pseudotumor diagnosis referred to the Department of Rheumatology of the Fray Antonio Alcalde Civil Hospital in Guadalajara, Jalisco, Mexico, from 2012-2018. We collected data that included demographics of the patient, symptoms, laboratory data that included antibodies, management plans and results.

Results: A total of 20 patients diagnosed with orbital pseudotumor, with a mean age of 42±18.5 years, 3 pediatric patients and 17% women. Clinical manifestations were: 90% unilateral, 90% lacrimal gland involvement, 75% ptosis/proptosis, 40% conjunctival hyperemia, 5% ocular pain, 20% decreased visual acuity, 15% headache and no optic nerve involvement. The findings were similar between adults and children. The most common diagnoses were: 40% idiopathic, 10% orbital cellulitis, 10% granulomatosis with polyangiitis and 5% each of the following: systemic lupus erythematosus, Sjögren’s syndrome, dacroyoadenitis and myositis due to IgG, Kawasaki’s disease, Mikulicz, meningioma and cavernous sinus aneurysm. All patients underwent excisional biopsy, and the histopathological report showed the following findings: 40% non-specific chronic inflammation, 30% non-specific chronic dacroyoadenitis, 5% granulomatous inflammation/vasculitis, 10% chronic sclerosing inflammation-IgG and 15% others. Only 9 of the 20 patients underwent immunological serology, finding positivity in: 15% for c-ANCA, 10% PR3, 5% p-ANCA, 10% MPO, 5% ANAs and 10% elevated blood levels of IgG4. Regarding treatment, 100% received glucocorticoids, and received immunomodulatory therapy: 20% received azathioprine, 5% mycophenolate mofetil, 20% methotrexate, 15% cyclophosphamide IV, 15% rituximab and 25% received no other medication.

Conclusion: The orbital pseudotumor might be the first manifestation of an autoimmune or autoinflammatory disease, the early and correct diagnosis is necessary to avoid permanent sequelae.

REFERENCES

Disclosure of Interests: None declared

AB1076
TREATMENT REVIEW OF ADULT-ONSET STILL’S DISEASE IN A TERTIARY HOSPITAL
Maria Dolores Arcila Durán, Clara Aguilera Cros, Lara Menendez, Isabel Madroñal García, Marina Gomez Vargas, Alberto Ruiz Román, Esteban Rubio Romero, Juan Povedano Gomez. Hospital Universitario Virgen del Rocío, Sevilla, Spain

Background: Adult-onset Still’s disease (AOSD) is a rare systemic inflammatory disorder of unknown aetiology, and approximately 60% or 70% of the patients can develop a chronic polyphasic form of the disease or a chronic polyarthritis. Due to the rarity of this disease, the treatment of AOSD is not based on a controlled study, but in the experience based on real cases.

Objectives: Describe the different treatments employed in a patient cohort diagnosed with adult-onset Still’s disease (AOSD).

Methods: Descriptive, retrospective study of patients treated in our Hospital (2008-2018), diagnosed with AOSD according to the classification criteria of Yamaguchi. The data were achieved by the review of the clinical records.

Results: Twenty-four patients (15 women), average age of 41±13 years, were included. Two women, with presentation on the symptoms at 8 and 3 years old, first diagnosed with systemic juvenile idiopathic arthritis (S-JIA), and then with AOSD.

The initial treatment was based in non-steroidal anti-inflammatory drugs (96%) and glucocorticoids (0.5-1 mg/kg/day) (96%) for symptom control, with the necessity to add oral or subcutaneous methotrexate at a dose of 15 mg per week in 13 patients (54%). Only two patients used acetyl salicylic acid as initial treatment, with no improvement.

<table>
<thead>
<tr>
<th>Previous treatment</th>
<th>Current treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Drug</td>
<td>n (%)</td>
</tr>
<tr>
<td>NSAIDs</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Glucocorticoids</td>
<td>23 (99%)</td>
</tr>
<tr>
<td>DMARDs</td>
<td>3 (12%)</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>3 (12%)</td>
</tr>
<tr>
<td>Leflunomide</td>
<td>3 (12%)</td>
</tr>
<tr>
<td>Hydroxychloroquine</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Sulphasalazine</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Biological treatments</td>
<td>3 (12%)</td>
</tr>
<tr>
<td>Tocilizumab</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Anakinra</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Rituximab</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Infliximab</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Baricitinib</td>
<td>2 (9%)</td>
</tr>
<tr>
<td>Etanercept</td>
<td>2 (9%)</td>
</tr>
</tbody>
</table>

Five patients used also biological treatments with a standard doses, with the necessity of various drugs to achieve their clinical remission. Nowadays, all patients have their clinical remission. Patient 1: Infliximab, rituximab, tocilizumab and baricitinib. Patient 2: Etanercept and rituximab. Patient 3: Etanercept, adalimumab and infliximab. Patient 4: Etanercept, infliximab and tocitumab. Patient 5: Infliximab, tocilizumab, baricitinib and sarilumab (good response to a anti-IL6, tocilizumab were removed because of a local reaction in the injection’s spot, although it had a good response too). Two patients with clinical remission with JAK-kinase inhibitors.