Research and possibly search for new parameters of the ILP activity are needed.

Disclosure of Interests: None declared


FR0589

IS THERE AN OVERLAP OF ANTEUNETRPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIDES WITH IGG4-RELATED DISEASE OR NOT?

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Background: Pseudotumor orbita, pachymeningitis, periarteritis could be seen in both ANCA-associated vasculitis and IgG4-RD. Sometimes it may be difficult to differentiate these two entities. The co-occurrence/concurrency of Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV) and IgG4-related disease (IgG4-RD) was recently published by a collaborative EUVAS group [1].

Objectives: Firstly, we aimed to investigate ANCA positivity of our IgG4-RD cohort. Secondly, a literature review of co-occurrence/concurrency of AAV and IgG4-RD was done.

Methods: Data of totally 62 patients with IgG4-RD in Hacettepe Vasculitis Center Database was used. Patients were diagnosed with IgG4-RD according to comprehensive diagnostic criteria [2]. Dataset of patients including demographic data, clinical characteristics, and imaging and laboratory findings of IgG4-RD was re-evaluated in terms of AAV and ANCA positivity.

At next step, we performed a systematic literature review of the PUBLISH database covering the time period until April 2018. Relevant publications were searched using the MeSH terms "IgG4-related disease and Eosinophil Granulomatosis with Polyangiitis", "IgG4-related disease and Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis" and "IgG4-related disease and Granulomatosis with Polyangiitis".

Results: Totally 29 (46.7%) of our patients had ANCA results. Out of 29 patients 15 (51.7%) were considered as probable, 10 (34.5%) as definite and 4 as possible (13.8%) for IgG4-RD. Three (10.3%) of these patients had ANCA positivity. All of these ANCA titers were in low degree positivity (MPO-ANCA 1/100, MPO-ANCA 1/32 and PR3 ANCA 1/100). These three patients didn’t have any findings of vasculitis and didn’t have granuloma in their biopsy. When we evaluate these three patients with regards to meeting the Ig G4- RD criteria, 1 was definite, 1 was probable and 1 was possible.

In literature review, we found 17 cases that having both features of IgG4-RD and AAV (Table). These cases were re-evaluated according to the ‘Comprehensive Diagnostic Criteria for IgG4-RD’. Diagnoses of IgG4-RD were definite in 11 cases (64.7%), probable in 2 cases (11.8%) and possible in 4 cases (23.5%). ANCA were positive in 15 of 17 patients (88%). ANCA were directed against proteinase 3 (PR3-ANCA) in 6 patients and were directed against myeloperoxidase (MPO-ANCA) in 5 patients. Other four cases had both MPO-ANCA and PR3-ANCA. All PR3-ANCA positive cases have high titer of ANCA, whereas only one MPO-ANCA positive case has high titer of ANCA.

Conclusion: None of our IgG4-RD patients have any overlap with ANCA-associated vasculitis. Only in 3 patients (10.3%), ANCA positivity was detected without any histopathologic evidence. Just two patients of literature review, seemed to be full compatible with both diseases. Even though ANCA-associated vasculitis and IgG4-RD share clinical features, we think this might be as co-occurrence instead of a histopathologic link.

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FR0590

UVETIS SECONDARY TO CHECKPOINT INHIBITORS

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Background: The introduction of immunotherapy (immune checkpoint inhibitors, ICI) has led to a revolution in oncological treatments. The inhibitors of CTLA4 (ipilimumab), of PD1 (pembrolizumab, nivolumab) and of the ligand of PD1 (atezolizumab, avelumab, durvalumab) regulate T activation and its effector function, being effective for the treatment of various types of cancer1. However, this effect leads to a series of immune-mediated adverse events, among which uveitis of autoimmune mechanism have been described in about 1% of treatments2, 3.

Objectives: Methods: descriptive study of retrospective review of the cases of a tertiary hospital with about 1000 treatments between the beginning of 2014 and the end of 2018.

Results: A series of 4 cases of uveitis of autoimmune origin associated with ICI is presented (see table). The series described has characteristics similar to the information previously reported in the literature2, with an incidence of around 0.4%, according to the previously described, with frequencies of ocular toxicity around 1%, being the uveitis is the most frequent form of presentation. The time of presentation of uveitis since the beginning of treatment has been in all cases in the first 6 months. The forms of presentation described before, ranges from anterior uveitis (AU) to panuveitis and often papilledema, usually bilateral, as are all those described in our series. In addition, 50% are AU (grade 2) and the other 50% panuveitis with papilledema (grade 3). In all the cases described, treatment was interrupted, a half it could be reintroduced without recurrence of uveitis. All the patients received topical treatment, the more severe systemic corticosteroids at doses of 0.5-1 mg/kg in accordance with the recommendations. The final prognosis was good, with preservation of VA in all cases, and topical and systemic treatment could be withdrawn in all patients. A patient treated with ipilimumab presented synechiae as a complication.

Disclosure of Interests: None declared

Conclusion: Uveitis is an infrequent, although potentially serious, immune-mediated side effect of ICI. Early recognition, discarding other causes of uveitis, particularly the masquerade syndrome, and early intervention are key to a good prognosis. The collaboration between the oncology teams and the ocular inflammation units must be close to establish the correct diagnosis and treatment, as well as to decide individually on the reintroduction or not of the oncological treatment. The implementation of registries on the adverse effects of these drugs can help to dimension the problem more accurately.

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