

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

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IS THERE AN OVERLAP OF ANTINEUTROPHIL 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/concurrence 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/concurrence 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 test.

At next step, we performed a systematic literature review of the PUBMED database covering the time period until April 2018. Relevant publications were searched using the MeSH terms "IgG4-related disease and Eosinophilic 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 titers of ANCA, whereas only one MPO-ANCA positive case has high titers 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.

REFERENCES:

- [1] Danlos, FX. *Autoimmun Rev*, 2017.
- [2] Umehara, H. *Mod Rheumatol*, 2012.

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Table: Distribution of Demographic, Clinical, Features in Seventeen Patients at Literature

Author	No. of Patients	Age (mean ± SD)	Gender (M/F)	ANCA	ANCA Titers	ANCA Specificity	ANCA Positivity	ANCA Titers	ANCA Specificity	ANCA Positivity	ANCA Titers	ANCA Specificity	ANCA Positivity	ANCA Titers	ANCA Specificity	ANCA Positivity	ANCA Titers	ANCA Specificity	ANCA Positivity
1	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
2	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
3	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
4	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
5	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
6	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
7	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
8	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
9	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
10	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
11	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
12	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
13	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
14	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
15	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
16	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive
17	1	64	M	PR3-ANCA	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive	1:100	PR3-ANCA	Positive

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UVEITIS 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.

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 synchiae as a complication.