AB1018

TREATMENT RESPONSE OF HYDRONEPHROSIS ASSOCIATED WITH IDIOPATHIC RETROPERITONEAL FIBROSIS, FOCUSING ON RATIO OF IGG4/IGG3 SERUM CONCENTRATION

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Background: Hydronephrosis, a common complication of idiopathic retroperitoneal fibrosis (iRPF), may lead to poor renal outcomes unless it is resolved. Pathological confirmation can help to identify the aetiology of the disease and determine the treatment strategy. But, in most cases, it is difficult to obtain sufficient tissue due to the location of fibrosis. In a recent study, parts of iRPF are correlated with IgG4-related disease characterised by elevated serum IgG4 levels (>135 mg/dL). Normal serum IgG3 level (21–176 mg/dL) has been known to be higher than normal serum IgG4 level (4–86 mg/dL). The reverse IgG4/IgG3 ratio has been suggested to be an IgG4-related disease component that distinguishes it from primary sclerosing cholangitis [1]. However, the ratio of IgG3 and IgG4 may be reversed in iRPF patients with hydronephrosis.

Objectives: We aimed to investigate the ratio of IgG subclasses as a predictive factor associated with treatment response of hydronephrosis in patients with iRPF.

Methods: We retrospectively recruited 19 iRPF patients with hydronephrosis who evaluated serum IgG subclasses in a tertiary hospital between 2004 and 2019. Hydronephrosis was evaluated on the basis of imaging findings. Medications and clinical and laboratory findings, including IgG subclasses, were reviewed following the diagnosis of hydronephrosis. Hydronephrosis improvement on subsequent images was evaluated to assess treatment response. Categorical data were compared using chi-square or Fisher's exact test. Continuous variables were compared using Mann-Whitney U test.

Results: At baseline, median serum IgG3 and IgG4 levels were 64 (IQR 37–82) mg/dL and 71 (IQR 40–171) mg/dL. Five patients had serum IgG4 levels > 135 mg/dL and 11 patients had the reverse serum level of IgG4/IgG3. On subsequent images (median follow-up at 3.2 [IQR 1.7–4.0] months), 11 patients showed hydronephrosis improvement. The proportions of positive ratio of serum IgG4/IgG3 (81.8% vs. 25%, $p = 0.024$), periaortic involvement (81.8% vs. 25%, $p = 0.024$) and high-dose glucocorticoid treatment (45.5% vs. 0%, $p = 0.045$) were significantly higher in patients with improvement than in those without improvement (Table 1). Interestingly, even in cases with normal serum IgG4 levels, patients with improvement showed a higher serum IgG4/IgG3 ratio than in those without improvement (median 1.5 vs. 0.7, $p = 0.038$).

Table 1. Clinical characteristics and treatment according to the short-term outcome of hydronephrosis

	Improvement (n = 11)	No improvement (n = 8)	p-value
Time to subsequent imaging (months) ^a	2.2 (1.3–4.2)	3.2 (2.0–3.8)	0.778
Males (n, %)	8 (72.7%)	7 (87.5%)	0.603
Periaortic involvement (n, %)	9 (81.8%)	2 (25%)	0.024
Impaired renal function (n, %) ^b	5 (45.5%)	2 (25%)	0.633
Serum IgG4 (mg/dL) ^a	114 (59–172)	43 (35–109)	0.152
Elevated serum IgG4 (n, %)	4 (36.4%)	1 (12.5%)	0.338
Serum IgG4/IgG3 ratio ^a	2.1 (1.2–4.9)	0.8 (0.4–1.0)	0.041
Positive ratio of serum IgG4/IgG3 (n, %)	9 (81.8%)	2 (25%)	0.024
Medical treatment (n, %)	7 (63.6%)	1 (12.5%)	0.059
High-dose glucocorticoid treatment (n, %)	5 (45.5%)	0 (0%)	0.045
Surgical intervention (n, %)	8 (72.7%)	4 (50%)	0.377

^a Values are median and interquartile range (25th–75th percentile)

^b Defined as serum creatinine level > 1.24 mg/dL

Conclusion: The reverse ratio of serum IgG4/IgG3 was associated with hydronephrosis treatment response, thus suggesting favourable responses to high-dose corticosteroid.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4159

AB1019

THERAPEUTIC PROFILE OF PATIENTS WITH VOGT KOYANAGI HARADA DISEASE FOLLOWED AT THE HOSPITAL DE CLÍNICAS. SAN LORENZO PARAGUAY.

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Background: Vogt-Koyanagi-Harada Disease (VKHD) is an autoimmune eye disease that causes chronic vision complications in young people. The treatment is based on corticosteroids and immunosuppressants, to which biological drugs are currently added. The visual prognosis depends on the stage in which it is diagnosed and the effective and timely treatment.^{1,2}

Objectives: Describe the evolution and the therapeutic profile of patients with VKHD who go to the Rheumatology Department of the Hospital de Clínicas. San Lorenzo Paraguay until November 2019

Methods: Descriptive cross-sectional study conducted at the Hospital de Clínicas, San Lorenzo Paraguay, where clinical histories of patients with this disease and who have regular follow-up were reviewed.

Results: 21 patients were included, 17 (81%) women, average age of 35 years. The diagnosis and treatment were made 15 (71%) in acute phase, 4 (19%) convalescent phase and 2 (10%) in chronic phase.

Corticosteroids: All received corticosteroids at diagnosis. Methylprednisolone IV 1g / day for 3 consecutive days in 8 (44%) cases and for 5 days in 10 (55.5%) cases. The first month everyone received prednisone orally at 1mg / kg / d on average (60–75 mg/d), at 3 months the average dose of prednisone was 37 mg / d, at 6 months of 23 mg/d and at 12 months 10 mg / d months.

Immunosuppressive treatment: They received 20 (95%) patients. Induction with monthly intravenous cyclophosphamide was performed in 8 (40%) cases and with Azathioprine in 12 (60%) cases. Of the patients treated with Azathioprine initially, in 5 (23%) patients it was necessary to switch to Cyclophosphamide IV due to failure or intolerance. The maintenance treatment was azathioprine in 13 (62%) of the patients, MTX in 4 (19%) and the association of both drugs was indicated in 3 (14%) patients. One case was refractory and received infliximab and subsequently Rituximab, which resulted in remission.

The best final Visual Acuity (AV) was recorded with the Snellen chart in 40 eyes (20 patients), 27 (67.5%) patients with good vision (20/20 to 20/50), 8 (20%) patients with decreased VA (20 / 50-20 / 200), and 5 (12.5%) with legal blindness (<20/200) and of these only 3 patients had unilateral involvement and one patient with bilateral blindness. Sequelae / complications: 6 (28.5%) patients presented cataracts and finally 3 (14%) glaucoma, 5 (24%) anterior / posterior synechiae.

Conclusion: The majority of patients in this series presented a good vision when the inflammatory process was resolved (AV 20 / 20-20 / 50), this probably related to early diagnosis and initial aggressive treatment with intravenous corticosteroid pulses and its association with immunosuppressants of entry as mentioned in the literature. The worst vision, (visual acuity <20/200) occurred in 5 of 40 eyes, corresponding to 4 patients, two of them diagnosed and treated in the chronic stage. Timely treatment improves visual prognosis.

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Acknowledgments: Department of Rheumatology

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.792

AB1020

APPLICATION OF THE 2019 ACR/EULAR CLASSIFICATION CRITERIA CONTRIBUTES TO THE DIAGNOSIS OF IGG4-RELATED DISEASE

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Background: IgG4-related disease (IgG4-RD) is a rare and systemic disease characterized by swelling or masses in the involved organs, elevated serum IgG4, marked IgG4+ plasma cells infiltration and fibrosis. However, lacking of pathological data limited the definite diagnosis of IgG4-RD. As results of high-quality clinical and epidemiological investigations collected, new classification criteria for