AB0610

PERSISTENT INFLAMMATION DESPITE TOCILIZUMAB TREATMENT IN 2 PATIENTS WITH LARGE VESSEL VASCULITIS AND AORTIC INVOLVEMENT REQUIRING AORTIC VALVE REPLACEMENT

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Background: Tocilizumab (TCZ) has recently been approved for treatment of giant cell arteritis (GCA). Despite clinical, radiologic and serologic improvement in the majority of patients, it remains unclear whether inflammation of the vascular wall in patients with large vessel vasculitis may persist.

Objectives: To report 2 patients who were treated with TCZ for large vessel vasculitis involving the ascending aorta. The patients underwent aortic valve replacement in both cases and partial replacement of the ascending aortic arch in one case.

Methods: Following therapy with glucocorticosteroids (GC) and TCZ, replacement of the aortic valve and of the partial ascending aorta were performed, followed by a histopathologic examination of the surgical specimen.

Results: We report two female patients, aged 40 and 67 years, both diagnosed with large vessel vasculitis involving the aortic arch. Aortitis of the ascending aorta and significant aortic insufficiency were demonstrated in both patients by PET-CT, MR angiography and transesophageal cardiac ultrasound.

In Pat 1, PET CT and MR angiography revealed vascular inflammation. Intravenous pulse GC was initiated, followed by oral prednisone with stepwise tapering and TCZ iv 8mg/kg/month. Two weeks after the 2nd infusion of TCZ, MR angiography showed significant reduction of gadolinium enhancement in the wall of the ascending aorta and the patient subsequently underwent valve replacement.

Pat 2 was diagnosed with vasculitis of the aortic arch by MR angiography. Therapy with oral GC (1mg prednisone/kg) and methotrexate was initiated. After 12 months, the treatment was switched to TCZ 162mg sc once weekly because of inadequate control of disease. The patient was monitored clinically and by PET CT and GCA was considered well controlled over 4 years. Due to continuous enlargement of the ascending aorta resulting in significant aortic insufficiency, the patient underwent replacement of the aortic valve and the ascending aorta.

Both patients showed normalization of CRP and ESR during TCZ treatment.

In summary, despite PET-CT and MR angiography showing reduction in vascular wall inflammation before surgery and CRP and ESR normalized, both patients showed histologically persistent lymphoplasmacellular infiltrates (predominantly CD3+CD4+ T cells in patient 1) and, in addition, giant cells in pat 2.

Conclusion: Despite adequate control of large vessel vasculitis involving the ascending aorta clinically as well as by imaging, the 2 patients reported here had to undergo valve replacement which showed persistent inflammation of the aortic wall despite treatment with TCZ.

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AB0611

A NEW SHAPE OF THE TREATMENT OF VASCULITIDES: SINGLE CENTER BIOLOGICAL AGENTS EXPERIENCE

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Background: Vasculitides are a group of inflammatory diseases with different clinical and pathological features of vital importance in various sized and types which may lead to organ failure and inflammation of the vascular wall. The framework for the treatment of vasculitis is similar to that used for many other systemic autoimmune rheumatic disorders, while the specific therapeutic regimen depends upon the nature and severity of the particular disorder (1,2).

Objectives: The aim of this study is to evaluate the efficacy of biological agents in 56 vasculitides patients with resistant/intolared to cDMARDs.

Methods: Between January 2015 and December 2018, fifty six patients who were diagnosed as vasculitis (Takayasu arteritis-TA, Granulomatosis with polyangiitis(GPA), Giant cell arteritis (GCA) and Behçet Syndrome (BS) at Sivas Cumhuriyet University Medical Faculty Rheumatology-Internal Medicine Department were included in to the study.

Results: 24 (43%) male and 32 (57%) female were included in the study. The median age of the patients was 56 years (min:19-max: 77) and the median age at diagnosis was 39 years (min 17-max 73). 14 (25%) patients with TA, 22 (39%) patients with GPA, 3 (5%) patients with GCA and 17 (31%) patients with BS included in this study. Tocilizumab treatment was used 6 of 8 patients who were resistant to anti-TNF. In 22 GPA patients with pulmonary and kidney involvement, Rituximab (RTX) treatment was started in 18 patients who were unresponsive to cDMARD and cyclophosphamide therapy. Plasmaexchange were used in 4 (18%) patients with alveolar hemorrhage. Indication of anti- TNF agents in behçet's syndrome were as follows panuveitis 70%, neuro-behçet 17.5% and vessel involvement 12.5% (Table 1). 1 GPA patient was died and other GPA, TA and BS patients have clinically response. Biological treatment is continued as maintenance therapy.

Conclusion: Biological treatments were used safely and effectively in vasculitis. In particular, RTX treatment is recommended for both induction and maintenance of GPA. Tocilizumab treatment is very effective in TA and GCA. However, large and long-term studies are needed in terms of long-term effects.

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Abstract AB0611 Table 1: Clinical Features Of Patients And Treatment Modalities

	n	Clinical features	cDMARDs	Anti-TNF	RTX
Takayasu	14 (25%)		6 (43%)	2 (14%)	6 (43%)
GPA	22 (39%)	Renal+pulmoner 18 (82%) Limited 4 (18%)	4 (18%)		18 (82%)
Behçet	17 (31%)	Uveitis 12 (70%) Neuro-behçet 3 (17.5%) Vessel inv. 2 (12.5%)		17 (31%)	
GCA	3 (5%)		2 (66%)	1 (33%)	

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AB0612

TEMPORAL ARTERY BIOPSYIN GIANT CELL ARTERITIS. INTERRELATION OF INFLAMMATORY PATTERNS WITH ANALYTICAL DATA AND CLINICAL MANIFESTATIONS

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Background: Giant cell arteritis (GCA) is a systemic vasculitis that affecting large and medium vessels, mainly the temporal artery and other arteries of extracranial localization (1).

Objectives: A series of 24 patients with clinical suspicion of GCA proved by a positive temporal artery biopsy (TAB) has been reviewed. Our aim was to asses a) histologic inflammatory patterns in the TAB and b) their possible correlation with analytical and clinical data.

Methods: Prospective study of 24 patients with positive TAB between January of 2016 until January of 2018. Clinical data, included: age, sex, symptomatology (cranial, visual or systemic manifestations) and physical examination of the temporal artery.

The analytical data registered were: ESR, CRP and haemoglobin. Furthermore, it has been considered if patients were receiving treatment with corticosteroids at the time of the biopsy.

Positive biopsies were classified into four categories: a) inflammation limited exclusively to the small vessels of the adventitia (SVV); b) vasa vasorum vasculitis (VVV); c) inflammation limited to the adventitia (ILA);

and d) transmural inflammation (TMI). The presence of giant cells, thrombosis and dystrophic calcification was also noted.

Results: We included a total of 24 patients with positive TAB: a) 19 patients (79.16%) presented TMI and b) 5 patients (20.84%) presented ILA. There were no cases with SVV or VVV.

No significant differences at baseline were observed between the 2 groups regarding sex and age, with a mean of 79.6 ± 7.2 years in TMI group and 78.8 ± 7.3 years in ILA group ($p = 0.83$).

There were no significant differences between the correlation of the clinical and analytical data and the 2 patterns of inflammation observed in the biopsy (**TABLE-1**).

Regarding the treatment at the time of performing the BAT, 16 (of 19) patients in the group with TMI (84.2%) and 4 (of 5) patients in the group with ILA (80%) were receiving treatment with oral glucocorticoid (Prednisone). No significant differences between the two groups were found in terms of treatment duration prior to BAT and nor in the dose received (**TABLE-2**).

Conclusion: In our sample, we didn't observe differences between the patterns of inflammation and the analytical and clinical data. It is important to recognise these patterns of inflammation in the TAB to avoid false negatives.

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TABLE-1. CLINICAL AND ANALYTICAL DATA.

Variables	TMI (n=19)	ILA (n=5)	P
Sex (women), n (%)	10 (52.6)	5 (100.0)	0.12
Age at disease onset, (mean $\pm$ SD)	79.6 ± 7.2	78.8 ± 7.3	0.83
CLINICAL DATA			
Any cranial symptoms	17 (89.5)	3 (60.0)	0.18
Headache	15 (78.9)	3 (60.0)	0.57
Scalp Tenderness	6 (31.6)	2 (40.0)	0.99
Abnormalities of temporal arteries	9 (47.4)	1 (20.0)	0.36
Jaw claudication	9 (47.4)	2 (40.0)	0.99
Visual loss	5 (23.8)	1 (20.0)	0.99
Any visual symptoms	15 (78.9)	3 (60.0)	0.62
Systemic signs/symptoms	11 (57.9)	4 (80.0)	0.51
PMR	5 (26.3)	3 (60.0)	0.29
Peripheral synovitis	0 (0.0)	0 (0.0)	0.99
ANALYTICAL DATA			
ESR (mean $\pm$ SD) (mm/H)	64.2 ± 36.7	74.0 ± 32.2	0.57
CRP (mean $\pm$ SD) (mg/dL)	10.2 ± 9.0	9.5 ± 8.8	0.75
Hemoglobin (mean $\pm$ SD) (g/dL)	11.6 ± 1.5	11.1 ± 2.4	0.43

TABLE-2. TREATMENT.

Variables	TMI (n=19)	ILA (n=5)	P
TREATMENT			
Patients taking Prednisone at the time of TAB	16 (84.2)	4 (80.0)	0.99
Prednisone dose at the time of TAB (median [IQR]) (mg/d)	40.0 [30.0-60.0]	270.0 [23.0-875.0]	0.44
Duration of Prednisone treatment before TAB (median [IQR]), days	5.0 [1.0-8.0]	3.0 [1.0-30.0]	0.90

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AB0613

PRESENTATION OF TAKAYASU ARTERITIS ACCORDING TO THE AGE OF ONSET IN A MONOCENTRIC ITALIAN COHORT

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Background: Takayasu arteritis (TA) is a large-vessel vasculitis usually affecting young women. The age of onset is though variable. Controversial data are reported about differences in disease features according to the age of onset.

Objectives: To compare clinical and vascular TA features according to TA age of onset.

Methods: We retrospectively evaluated onset demographic and disease features in TA patients followed-up at our Hospital between January 2000 and August 2018. We divided our cohort into 3 groups according to the age of onset: ≤ 20 (Group 1), 21-40 (Group 2), > 40 (Group 3) years. Diagnostic delay (DD) was defined as the time interval between the first TA symptom and TA diagnosis. Non parametric statistic tests were used.

Results: 126 patients (12 male, 114 female) were analyzed. Mean age at onset was 31.46 ± 13.38 years. Group 1 and 3 included 30 (23.81%) patients each, Group 2 66 (52.38%). Mean age at onset was 16.23 ± 3.20 years in Group 1, 29.59 ± 5.80 in Group 2 and 50.8 ± 7.16 in Group 3. Mean DD was different between the 3 groups: 117.63 ± 129.79 months in Group 1, 56.47 ± 76.98 in Group 2 and 30.80 ± 40.53 in Group 3 (Group 1 vs 2, $p=0.021$; Group 1 vs 3, $p<0.001$; Group 2 vs 3, $p=0.089$). Clinical and laboratory features at TA onset are summarized in Table 1. Among all disease features only arterial hypertension was statistically more prevalent in Group 3 vs Group 1 and 2 (56.67% vs 36.67% and 30.30% respectively, $p=0.048$). When evaluating vascular involvement, abdominal aorta was more frequently affected in Group 1 (63.33% vs 34.85% and 56.67%, $p=0.018$) while right iliac and left iliac arteries in Group 3 (40% vs 23.33% and 10.61%, $p=0.004$; 36.67% vs 23.33% and 12.12%, $p=0.024$ respectively).

Abstract AB0613 Table 1

Clinical and laboratory features	Group 1 n 30 (%)	Group 2 n 66 (%)	Group 3 n 30 (%)	p-value
Systemic symptoms	21 (70)	37 (56.06)	15 (50)	0.254
Carotidynia	11 (36.67)	12 (18.18)	5 (16.67)	0.116
Lower extremities claudication	7 (23.33)	18 (27.27)	4 (13.33)	0.363
Upper extremities claudication	10 (33.33)	31 (46.97)	7 (23.33)	0.083
Δ blood pressure	10 (33.33)	16 (24.24)	6 (20)	0.526
Reduced/absent pulse	18 (60)	29 (43.94)	11 (36.67)	0.178
Bruits	15 (50)	20 (30.30)	8 (26.67)	0.114
Angina abdominis	6 (20)	6 (9.09)	3 (10)	0.376
Myocardial infarction	0	5 (7.58)	4 (13.33)	0.154
Heart failure	0	6 (9.09)	0	0.078
Arterial hypertension	11 (36.67)	20 (30.30)	17 (56.67)	0.048*
Headache	10 (33.33)	9 (13.64)	8 (26.67)	0.067
Syncope	9 (30)	9 (13.64)	6 (20)	0.180
Stroke	2 (6.67)	6 (9.09)	2 (6.67)	1
Amaurosis	4 (13.33)	4 (6.06)	3 (10)	0.499
$\uparrow$ inflammatory markers	25 (83.33)	52 (78.79)	25 (83.33)	0.869
Vascular surgery	8 (26.67)	13 (19.70)	11 (36.67)	0.202

Abstract AB0613 Table 2

Artery	Group 1 n 30 (%)	Group 2 n 66 (%)	Group 3 n 30 (%)	p-value
Coronary	2 (6.67)	7 (10.61)	4 (13.33)	0.274
Pulmonary	5 (16.67)	12 (18.18)	1 (3.33)	0.146
R subclavian	14 (46.67)	32 (48.48)	17 (56.67)	0.771
L subclavian	18 (60)	43 (65.15)	22 (73.33)	0.554
R CC	19 (63.33)	36 (54.55)	18 (60)	0.701
L CC	21 (70)	38 (57.58)	19 (63.33)	0.497
R vertebral	3 (10)	11 (16.67)	3 (10)	0.648
L vertebral	4 (13.33)	12 (18.18)	6 (20)	0.863
Brachiocephalic	10 (33.33)	22 (33.33)	6 (20)	0.425
Ascending ao	11 (36.67)	19 (28.79)	6 (20)	0.369
Arch	9 (30)	20 (30.30)	11 (36.67)	0.821
Descending ao	10 (33.33)	17 (25.76)	9 (30)	0.756
Abdominal ao	19 (63.33)	23 (34.85)	17 (56.67)	0.018*
Celiac	7 (23.33)	23 (34.85)	12 (40)	0.344
Superior mesenteric	8 (26.67)	22 (33.33)	6 (20)	0.398
R renal	9 (30)	16 (24.24)	4 (13.33)	0.297
L renal	7 (23.33)	15 (22.73)	5 (16.67)	0.843
R iliac	7 (23.33)	7 (10.61)	12 (40)	0.004*
L iliac	7 (23.33)	8 (12.12)	11 (36.67)	0.024*

Abbreviations: right (R), left (L), common carotid (CC), aorta (ao)