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AB0650

THROMBOTIC MICROANGIOPATHY ASSOCIATED TO ANCA-POSITIVES VASCULITIS: A FRENCH RETROSPECTIVE CASE CONTROL STUDY AND LITERATURE REVIEW

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Objectives: In this large nationwide French case-control study, we describe the features of TTP and ANCA-positive vasculitis; compare to vasculitis without associated TTP; describe the outcome and treatments.

Methods: We collected all cases with TTP and associated vasculitis. We conducted a literature review using PubMed, Web of Science, congress posters from January 2005 to August 2017 of PTT and ANCA-positive vasculitis.

A control group of MPA without TTP during all the disease follow-up was extracted from the Saint Antoine and Montfermeil Hospital patients with vasculitis. Firstly we compared our PTT cases and the literature review cases and secondary with a control group ANCA vasculitis without MAT.

Results: 8 patients with MAT secondary to ANCA associated vasculitides were included in our French series 75% of Women with a median age 45 years, 21–76 positive ANCA in 50% and 37.5% is MPO, BVSA score at 16, FFS at 1. In 10 literature cases 90% Women, a median age 60 years [17–77], positive ANCA in 100% and 30% is MPO, BVSA score at 39, FFS at 1.

The clinical features at the diagnosis of vasculitis were fever (n=5; 62%), ENT involvement (25%), kidney crescentic glomerular involvement (n=6; 75%) with kidney failure in joint involvement (n=2; 25%) polyarthralgia type, with gastrointestinal involvement type mesenteric ischemia and pericarditis and lung involvement (nodules and alveolar haemorrhage) (n=1; 12.5%), in one case each Median C-reactive protein levels were at 49 mg/L [1–204], with creatininemia at 170 mg/dl [80–588]. ANCA were present in 4 patients (50%), MPO in 3 cases (37.5%). The time between the diagnosis of vasculitis and TTP was 9 months [0–51].

TTP features were: Hb at 7.8 g/dl [4.8–10], LDH 1658 [777–3110], platelets 33000 [3000–1 25 000], haptoglobin at 0 [0–0.05], creatinine at 205 [80–757]. ADAMTS 13 levels were at 10 [1–138], and x patients have normal ADAMTS13 levels, with anti ADAMTS 13 antibodies in 1 case. Plasma exchanges were done in all patients with median of 10 exchanges [4–23], 37.5% are dialyzed.

Conclusions: In comparison to our cases, the literature patients have similar organs involvement, but median creatininemia levels and BVAS levels were higher in the literature cases. Considering TTP features, our cases have less frequent active vasculitis, less important creatinine levels and thus less recurs to kidney dialysis.

In vasculitis associated with TTP, there was no significant differences in organ involvements, BVAS and FFS scales values, laboratory data and ANCA levels. Only creatininemia as expected was higher in vasculitis associated with TTP (225 Mmol vs 150 Mmol, p=0.0044).

Disclosure of Interest: None declared

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AB0651

FREQUENCY OF PULMONARY HYPERTENSION IN BEHÇET'S DISEASE

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Background: Behçet's disease (BD) is a systemic vasculitis that involvement of pulmonary arteries can be seen.

Objectives: The aim of this study was to determine the frequency and the causes of pulmonary hypertension (PH) in patients with BD.

Methods: We studied consecutively 154 BD patients who were fulfilled the International Study Group criteria for diagnosis of BD. All patients were evaluated with transthoracic echocardiography (TTE) for the presence of PH. BD patients were categorised according to the involved organs in 5 groups: group 1 mucocutaneous and articular, group 2 ocular, group 3 vascular, group 4 gastrointestinal and group 5 neurologic involvements. The presence of PH was defined as estimated sPAB ≥40 mmHg, by TTE. Every subject evaluated by a detailed medical history and

physical examination was performed. Additional laboratory results were obtained from hospital file records.

Results: The mean age (SD) and the median (min-max) disease duration of the patients were 41.8±12.6 years and 126 (6–540) months respectively. PH was detected in 17 (11%) BD patients. Only 9 (52%) patients were symptomatic (NYHA FC >1). Left sided heart disease (Group II: 9 (52%) patients) was the leading cause of PH. Four (23%) patients had group IV PH and 75% (3/4) were symptomatic. Diastolic dysfunction (DD) was found in 32 (20.8%) patients and only 1 patient had systolic dysfunction. The number of patients with DD was significantly higher in patients with PH as compare to patients without PH (8 (47.1%) vs 24 (17.6%), p=0.005). There were no difference in demographic and clinical features of patients with and without PH. Only acneiform lesion was more frequently in patients without PH as compare to patients with PH, p=0.047 (table 1). There were no differences in frequency of PH in BD groups (table 2).

Abstract AB0651 – Table 1. Demographic and clinical characteristics features of Behçet disease

	All	sPAP ≥40 mm HG	sPAP<40 mm HG
Female, n (%)	62 (40.3)	6 (35.3)	56 (40.9)
Age, mean, SD	41.8±12.6	48.1±14.6	41.0±12.1
Disease duration (month), median (min-max)	126 (6–540)	168 (12–540)	120 (6–480)
Smoking, n (%)	76 (49.4)	7 (41.2)	69 (50.4)
Never smoker	47 (30.5)	5 (29.4)	42 (30.7)
Current smoker	31 (20.1)	5 (29.4)	26 (19.0)
Ex-smoker			
Oral ulcer, n (%)	154 (100)	17 (100)	137 (100)
Genital ulcer, n (%)	104 (67.5)	11 (64.7)	93 (67.9)
EN, n (%)	64 (41.6)	7 (41.2)	57 (41.6)
Papulo-pustular lesion, n (%)	35 (22.7)	3 (22.7)	32 (23.4)
Acneiform lesion*, n (%)	105 (68.2)	8 (47.1)*	97 (70.3)*
Articular, n (%)	33 (22.7)	4 (23.5)	31 (22.6)
Uveitis, n (%)	75 (48.7)	9 (52.9)	66 (48.2)
Pathergy, n (%)	40 (26)	6 (35.3)	34 (24.8)
Vascular, n (%)	48 (31.2)	9 (52.9)	23 (31.1)
Neurologic, n (%)	18 (11.7)	2 (11.8)	16 (11.7)
Gastrointestinal, n (%)	12 (7.8)	1 (5.9)	11 (8.0)

sPAP: Systolic pulmonary artery pressure, EN: Erythema nodosum

Abstract AB0651 – Table 2. Behçet disease group distribution in the pulmonary hypertension patients

Behçet disease (n)	Mucocutaneous and articular (39)	Ocular (75)	Vascular (48)	Gastrointestinal (12)	Neurologic (18)	p
sPAP ≥40, n (%)	2 (5.1)	9 (12)	9 (18.8)	1 (8.3)	2 (11.1)	0.413

sPAP: Systolic pulmonary artery pressure

Conclusions: The most frequent cause of PH in BD patients was group II PH. There were also significant number of patients with group IV PH. Pulmonary endarterectomy may be an option of treatment in inactive patients with group IV PH.

Disclosure of Interest: None declared

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AB0652

PET/MR IN LARGE-VESSEL VASCULITIS: CLINICAL VALUE FOR THE DIAGNOSIS AND ASSESSMENT OF DISEASE ACTIVITY

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Background: The diagnosis and the activity determination could be challenging in large-vessel vasculitis (LVV).

Objectives: The aim of this study was to analyze the value of hybrid PET/MR in LVV.

Methods: All consecutive patients with LVV who underwent PET/MR were included. PET/MR patterns were defined as inflammatory in the case of positive PET (grade=3) and abnormal MR (stenosis and/or wall thickening) and fibrous in the case of negative PET (grade 1 or 2) and abnormal MR.

Results: Thirteen patients with median age at 67 years (23–87 years) and 10 (77%) females were included, and underwent 18 PET/MR scans. Eleven PET/MR performed at diagnosis (n=4) or relapse (n=7) and 7 in patients in remission. 8/18 (44%) had PET/MR inflammatory pattern and 3/18 (17%) had fibrous pattern. PET/MR were normal in 2/10 (20%) cases of TA versus 5/8 (62%) cases of GCA

($p=0.3$). The median SUVmax was 3.0 [1.8–8.6] without significant difference between GCA and TA: 3.4 (2.1–8.6) versus 2.6 (1.8–7.1) ($p=0.4$), respectively. Eleven PET (61%) were performed under treatment, which consisted of steroids with a median dose at 30 mg/day [3–240]. Among 11 patients with active disease, 8 had inflammatory patterns and 3 had normal PET/MR, i.e. a sensibility of 73%, and the sensibility increased to 100% in patients with active TA disease. Median SUVmax were 4.7 [2.1–8.6] in patients with active disease versus 2 [1.8–2.6] in patients with remission ($p=0.003$).

Conclusions: PET/MR is a new hybrid modality of imaging which is interesting for the diagnosis and the follow-up of large-vessel vasculitis.

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AB0653 PREDICTION OF RELAPSES IN AUTOIMMUNE LARGE-VESSEL VASCULITIS – TOWARDS PERSONALISED IMMUNOSUPPRESSIVE TREATMENT STEWARDSHIP

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Background: Giant cell arteritis (GCA) is an autoimmune disease of the large arteries. Treatment consists in long-term immunosuppression with glucocorticoids (GC). About half of the patients have disease flares ('relapses') despite standard therapy. Tocilizumab (TCZ) – an anti-IL6 receptor antibody – is highly effective in reducing relapses, but has high costs.

Objectives: Here, we tested whether the initial clinical presentation and/or immunological findings might predict a GCA patient subset with poor response to GC. These might benefit from early TCZ therapy.

Methods: We performed a chart review on 113 patients from our prospective GCA cohort over the first two years after diagnosis. All had a follow-up of at least three months (median follow-up 24 months, IQR 12.6–24). Clinical findings at diagnosis, routine labs (at 0, 1, 3, 6, and 12 months) and therapy information (drug and dose) were extracted from the electronic database.

Relapses were defined as the presence of GCA-related symptoms (ischaemic pain, polymyalgia (PMR)) and/or elevated systemic inflammation parameters (CRP, ESR) that responded to an increase in GC-dose. GC receptor (GCR) expression levels in T cells were assessed using flow cytometry. Patients were genotyped for two polymorphisms in the Glucocorticoid receptor gene (NR3C1) that have been associated with steroid-responsiveness in other autoimmune disease (Systemic lupus, Pemphigus...).

Results: Over the first 12 months, 50.6% experienced at least one relapse. The majority of relapses occurred after three months of treatment (median time to relapse 102 days, range 19–312). This is when the GC dose is tapered below 20 mg/d. 'Relapsers' had an average of 1.66 (range 1–4) relapses in the first year. Patients with fever at initial presentation had a 2.2-fold (CI 1.1–5.05) higher risk to experience relapses ($p=0.02$). Other clinical findings were not associated with subsequent relapses. Low lymphocytes after the first month of therapy was the only lab value associated with relapse free follow up (698/ul vs 1021/ul, $p=0.02$). This was independent of the cumulative GC dose that 'non-relapsers' and

'relapsers' received in this period (1747 mg vs. 1710 mg mg, $p=0.4$). Relapsers had lower GCR expression levels, as assessed by flow cytometry.

Conclusions: Fever, lack of lymphocytopenia after one month of therapy and low GCR expression are risk factors for relapses in GCA. Low GCR expression combined with absence of lymphocytopenia during high dose GC therapy points at a constitutional steroid-resistance in relapsers. Whether patient stratification based on these parameters allows to safely adapt ('personalise') the intensity and/or duration of GCA treatment needs to be tested in a prospective clinical trial.

Disclosure of Interest: None declared

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AB0654 CLINICAL PROFILE AND RISK FACTORS OF INFECTIONS IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS (AAV) – 18-YEAR DATA FROM A SINGLE TERTIARY CENTRE

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Background: Despite advances in the treatment of AAV, there are still considerable morbidities related to treatment-related complications. Infection is one of the most commonly encountered problems in patients on immunosuppressive therapy.

Objectives: This single centre, retrospective study reviewed the clinical features and investigated the risk factors for infections among patients with AAV.

Methods: 104 patients with AAV diagnosed between January 2000 and December 2017 in a tertiary hospital were included. Demographic data and clinical parameters were reviewed. Logistic regression was performed to identify factors predicting infections.

Results: Around two-thirds of the 104 patients included were female (63.5% $n=66$). Mean age at diagnosis was 64.4-year-old. The majority (65.4%, $n=68$) had microscopic polyangiitis (MPA), 19.2% ($n=20$) had granulomatosis with polyangiitis (GPA) and 15.4% ($n=16$) had eosinophilic granulomatosis with polyangiitis (EGPA).

More than half of the patients (58%, $n=61$) experienced at least one episode of infection and 22% ($n=23$) had recurrent sepsis during their disease course. Infection was the leading cause of mortality of the 56 deceased patients in this series. Infections were less frequent in EGPA patients compared to their counterparts with MPA and GPA (37.5% vs 61.8%–65%).

Most infections were bacterial and multiple-drug resistant organisms were the causative agents in 8 patients. Two had neutropenic sepsis. Three had M. tuberculosis and five had herpes zoster. One had concomitant VZV and pneumocystis jiroveci pneumonitis.

Abstract AB0654 – Table 1.

Factors predicting infection	Odds Ratio ($p<0.05$)
– Age	1.034
– Renal insufficiency (serum creatinine ≥ 140 $\mu\text{mol/L}$) at diagnosis	5.452
– Disease-related organ failure	3.006
– Dialysis support	6.504
– Use of cyclophosphamide as induction agent	2.956

Conclusions: Infections were common and often led to significant morbidities and mortalities among AAV patients. Risk factors included age, renal insufficiency (serum creatinine >140 $\mu\text{mol/L}$) on presentation, disease-related organ failure, need for renal supportive therapy, and the use of cyclophosphamide as induction agent.

Disclosure of Interest: None declared

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AB0655 AGREEMENT BETWEEN 18-FDG PET/CT AND CLINIMETRIC TAKAYASU ACTIVITY SCORES

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Background: The 18-FDG PET/CT is an objective tool employed for the diagnosis of Takayasu arteritis and also is used for the assessment of disease activity of this vasculitis.¹ There are few clinimetric scores developed for Takayasu clinical activity assessment such as the Indian Takayasu Clinical Activity Score (ITAS2010/ITAS.A),² the National Institutes of Health criteria (NIH score) from USA³ and the Mexican effort by Dabague-Reyes (DR score).⁴ The validity and