

dacryoadenitis (26 patients) and myositis (3 patients). Patients with orbital mass (group 1) and dacryoadenitis (group 2) were included into further investigation.

Results: Patients in group 1 had systemic disease in 51,1% compared to 7,7% in group 2. Patients with orbital mass presented with more severe clinical manifestations, including significant exophthalmos (82,2%), periorbital oedema (95,5%) and hyperemia (46,7%), strabismus (31,1%), globe movement restriction (71,1%), chemosis (8,9%). Orbital mass was accompanied by necrotizing scleritis in 7 and PUK in 6 patients. Symptoms included orbital pain (88,9%) and binocular diplopia (73,3%). BCVA in this group was 0.67. Clinical outcome was relatively unfavourable: in 24,4% BCVA was less than 0.3, in 5 patients enucleation was performed, in 24,4% strabismus developed and in 73,3% globe movement restriction occurred. Clinical course and outcome were significantly more favourable in patients with dacryoadenitis. Periorbital oedema, exophthalmos and chemosis did not show significant difference in groups, though orbital pain, binocular diplopia, globe movement restriction, strabismus, optic neuropathy, necrotizing scleritis, PUK were observed in group1 significantly more often. Clinical outcome in group 2 was much more favourable with no cases of enucleation and persistent visual loss. They only had moderate exophthalmos 1–3 mm (65,4%), strabismus (7,7%) and mild globe movement restriction (19,2%).

Conclusions: Three types of orbital involvement in GPA were proposed: orbital mass, dacryoadenitis and myositis. Patients with orbital mass are characterised more often with systemic disease, more severe clinical course not uncommon associated with other ocular manifestations (necrotizing scleritis, PUK, orbital walls destruction), relatively unfavourable outcome with high level of morbidity (optic nerve atrophy, anophthalmos, strabismus) and higher rate of recurrences.

Disclosure of Interest: None declared

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AB0659

PREVALENCE AND CLINICAL FEATURES OF CRANIAL AND EXTRACRANIAL GIANT CELL ARTERITIS

E. Fernández¹, I. Monjo¹, L. Coronel², D. Benavent¹, A. Balsa¹, E. De Miguel¹.

¹Rheumatology, Hospital Universitario La Paz, Madrid; ²Rheumatology, Hospital Universitario Nuestra Señora de La Candelaria, Santa Cruz de Tenerife, Spain

Background: Giant cell arteritis (GCA) is the most common systemic vasculitis in adults. Although it typically affects the cranial branches derived from the aortic arch, there is increasing evidence of the damage occurring to large vessel extracranial arteries, which is usually misdiagnosed. This subset of large vessel extracranial GCA may have specific clinical features that could hinder the diagnosis, which highlights the need for a different treatment and follow up.

Objectives: To analyse the frequency and clinical and analytical features in cranial and extra-cranial GCA.

Methods: An observational retrospective descriptive study analysing data from patients with GCA was performed in our hospital. Colour duplex sonography (CDS) studies carried out in the last 29 months were revised. Standardised CDS images from the frontal and parietal branches of the temporal superficial artery (TA) and axillary artery (AXE) with GCA compatible image with intra or extra-cranial involvement (hypochoic, homogeneous and circumferential thickening of the artery wall ≥ 0.34 mm in TA, and ≥ 1 mm in AXE, respectively). SPSS version 23 were used to analyse a total of 29 variables.

Results: Out of the 119 patients that were tested in our fast track GCA clinic, with standardised CDS of TA and AXE, 22 had GCA. All GCA patients was halo sign in CDS. From them, 7 had large vessel involvement (31,8%), from whom 6 were mixed and 1 had exclusively extra-cranial involvement. Mean age at diagnosis was 76 ± 9.7 SD in the extracranial GCA arteritis and 79 ± 5.7 SD in the cranial GCA. The extracranial subset had a greater number of men than the cranial (28,6% and 26,6%, respectively) and suffered usually more fever (28,5% > 6,6%, respectively), constitutional syndrome (85,7% > 40%, respectively) or polymyalgia rheumatica (42,8% > 40%, respectively). However, they suffered with more frequency from headache (85,7% vs 93,3%) and jaw claudication (28,5% vs 33,3%). None of them suffered from visual impairment or central nervous system involvement; both do appeared in the cranial GCA group in 13,3% and 6,6%. In the blood tests, mean \pm standard deviation was: ESR 87.6 ± 36 mm/h in cranial GCA and 89.5 ± 19.5 SD mm/h in extracranial GCA, CRP 65 ± 57.6 and 82.6 ± 48.4 mg/L and Hb 11.4 ± 1.3 and 12.1 ± 1.3 g/dl, respectively. Patients with large vessel involvement met ACR criteria in 80% opposite 92,8% from these with solely cranial GCA. AT biopsy was performed in 7 patients in the cranial subset and 2 in the extra-cranial, testing positive for GCA in 5 (71,4%) in the first group, and none in the second one. CT-PET was performed in 2 patients with cranial GCA with negative results, and in 4 with large vessel involvement, testing positive in 50%. Despite the large differences, the statistical analysis didn't reach statistical significance due to the small sample size.

Conclusions: One third of the patients in our study had large vessel involvement, making axillary vessel CDS an important tool for the clinical exam of this disease. These patients tend to be younger and start more often with fever or general

syndrome and less with GCA typical symptoms like headache, jaw claudication or visual loss.

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AB0660

THE ROLE OF ANTI-NEUTROPHIL CYTOPLASMIC AUTOANTIBODY SPECIFICITY FOR MPO OR PR3 IN PHENOTYPE OF ANCA ASSOCIATED VASCULITIS: KNOWING THE AUTOIMMUNITY IN LATIN AMERICA

E. Calle¹, M. Calle², H.B. Cabrera³, D. Jaramillo-Arroyave^{1,4,5}, A.L. Vanegas-García^{6,7}, G. Vásquez-Duque¹, M. Restrepo-Escobar¹, L.A. González-Naranjo¹, J. Hernández-Zapata¹, C.H. Muñoz-Vahos^{1,5,7}. ¹Grupo de Reumatología Universidad de Antioquia, Medellín; ²Universidad CES, Medellín; ³Medicina Interna, Universidad de Antioquia; ⁴Universidad CES; ⁵IPS Universitaria Clínica León XIII, Medellín; ⁶Grupo de Reumatología Universidad de Antioquia, Medellín; ⁷Hospital Universitario San Vicente Fundación, Medellín, Colombia

Background: Antineutrophil cytoplasmic antibodies (ANCA) are present in up to 90% granulomatosis with polyangiitis, 80% microscopic polyangiitis and 70% eosinophilic granulomatosis with polyangiitis. MPO-ANCA has been associated with vasculitis limited to the kidney, chronic renal damage and less frequent gastrointestinal or respiratory tract involvement. PR3-ANCA are characterised by destructive lesions of the ears, nose and throat, alveolar haemorrhage, combination of upper and/or lower respiratory tract involvement with renal compromise and increased number of relapses. The frequency of pulmonary involvement is similar in both serotypes, and most ANCA associated vasculitis (AAV) patients are diagnosed between ages 50 and 70 years.

Objectives: To describe differences in clinical profiles of patients with AAV regarding ANCA specificity against MPO or PR3 in a Colombian based adult population

Methods: All medical records of patients with a diagnosis of AAV in two high complexity hospitals in Medellín, Colombia from January 1, 2014 to December 31, 2016 were reviewed. The clinical and demographic characteristics were abstracted and analysed with descriptive and inferential statistics in SPSS.22

Results: Of 59 cases of AAV, 44 were positive for MPO or PR3-ANCA with male predominance (65,5% men vs 34,5% women) and similar age at diagnosis (47 years in MPO-ANCA vs 50 in PR3-ANCA). MPO-ANCA group had more fever and weight loss (34,8% vs 20%), arterial hypertension (34,5% vs 26,7%), hematuria (34,5% vs 26,7%), proteinuria (31% vs 26,7%), creatinin higher than 5.6 mg/dL (20,7 vs 13,3%), myalgias (13,8 vs 0%) pachymeningitis (7% vs 0%) and skin compromise. PR3-ANCA patients had more arthralgias/arthritis (40% vs 31%), escleritis (33% vs 13,8%), episcleritis (13,3% vs 0%) and uveitis (10% vs 7%).

Conclusions: In this Latin American population ANCA specificity affected the phenotype of clinical disease. MPO-ANCA patients had more constitutional symptoms, renal and central nervous system compromise while PR3-ANCA patients showed more articular and ocular involvement.

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AB0661

ASSESSMENT OF DAMAGE AND PROGNOSIS IN PATIENTS WITH ADULT IGA VASCULITIS: RETROSPECTIVE MULTICENTERED COHORT STUDY

E. Alibaz-Oner¹, A. Omma², A. Sari³, Ü. Gazel¹, D. Üsküdar Cansu⁴, A. Yazıcı⁵, A. Cefle⁵, C. Bes⁶, Ö. Karadağ³, H. Direskeneli¹. ¹Rheumatology, Marmara University, School of Medicine, Istanbul; ²Rheumatology, Ankara Numune Research and Training Hospital; ³Rheumatology, Hacettepe University, School of Medicine, Ankara; ⁴Rheumatology, Osmangazi University, School of Medicine, Eskişehir; ⁵Rheumatology, Kocaeli University, School of Medicine, Kocaeli; ⁶Rheumatology, Bakırköy Sadi Konuk Research and Training Hospital, Istanbul, Turkey

Objectives: IgA Vasculitis is a leukocytoclastic vasculitis involving small vessels with depositions of immune complexes containing IgA. IgA Vasculitis is a predominantly paediatric vasculitis. There is limited data for the prognosis of adult IgA Vasculitis, with also no damage assessment. In this study, we aimed to evaluate

the clinical characteristics, treatment, outcome and damage of patients with adult IgA Vasculitis.

Methods: We assembled a retrospective cohort of patients with adult IgA Vasculitis from tertiary Rheumatology Centres in Turkey. The demographics, clinical characteristics, treatment and outcomes of patients were abstracted from medical records. Birmingham Vasculitis Activity Score (BVAS), prognostic Five Factor Score (FFS) and vasculitis damage index (VDI) were calculated.

Results: The study included 103 (male/female: 67/36) patients with adult IgA Vasculitis. The mean age was 42.6±17 years. Infection history within 6 weeks before presentation was present in 40 (38.8%) patients (32 upper respiratory tract, 3 urinary tract, 2 gastrointestinal, 3 others). Cutaneous manifestations and arthritis/arthralgia were the most common clinical manifestations (table 1). 92 (89.3%) patients were treated with oral glucocorticoids (GC). Pulse GC treatment was also given to 29 (28.1%) patients. As additional immunosuppressive agents, azathioprine was given to 36 (34.9%) and pulse cyclophosphamide to 13 (12.6%) patients. Fifty-nine patients (58.2%) had follow-up of mean 35.6 months. Eleven (18.6%) patients relapsed during follow-up. While 5 relapses were major, six of them were minor relapses. At the last visit, disease status was evaluated as active or treatment failure by the treating physician in 7 (11.8%) patients. The rate of chronic renal failure was 8.3%(n=5). Mortality was 1.6% (n=1) during follow-up, due to pneumonia. The mean VDI score was 0.3 in the last visit. Twelve (20.3%) patients had at least one damage item at the end of follow-up period.

Abstract AB0661 – Table 1. Baseline clinical characteristics of patients with adult Henoch Schönlein Purpura

Adult Henoch Schönlein Purpura (n=103)	
Laboratory parameters	
Anemia (<12 mg/dl for female, <13 mg/dl for male) (n, %)	36 (35%)
Erythrocyte Sedimentation Rate (mm/hour) *	34.7 ± 22
C-reactive protein (mg/l)†	18 (1.297)
Proteinuria (>300mg/24 hours)	47 (45.6%)
Creatinine (mg/dl)†	0.8±0.3
Hepatitis B positivity (n)	8/97 (8.2%)
Hepatitis C positivity (n)	0/97
ANA positivity	21/97 (21.7%)
RF Positivity	5/88 (5.7%)
c-ANCA positivity	2/97 (2.1%)
p-ANCA positivity	1/95 (1.1%)
Clinical Manifestations, n/103 (%)	
Fever	24 (23.3%)
Weight loss	26 (25.2%)
Myalgia/Weakness/Leg tenderness	44 (42.7%)
Arthritis and/or arthralgia	87 (84.5%)
Neurologic manifestations	1 (0.9%)
Testicular pain or tenderness	4 (3.9%)
Recent onset or severe hypertension	2 (1.9%)
Cutaneous Manifestations	97 (94.2%)
Peripheral limb edema	14 (13.6%)
Gastrointestinal manifestations	71 (68.9%)
Cardiac manifestations	5 (4.8%)
Ocular manifestations	3 (2.9%)
Pulmonary manifestations	3 (2.9%)
FFS=0	71 (68.9%)
FFS=1	24 (23.3%)
FFS=2	5 (4.9%)
FFS=3	3 (2.9%)
BVAS score at diagnosis*	6±3

ANA: Anti nuclear antibody, FR: Rheumatoid factor, ANCA: Antineutrophilic cytoplasmic antibody, FFS: Five Factor Score, BVAS: Birmingham Vasculitis Activity score. *Mean ±SD †Median (Minimum-maximum)

Conclusions: Our results showed that approximately one fifth of patients with adult IgA Vasculitis had relapses during follow-up and had at least one damage item at the end of follow-up. Although, 31% of patients had FFS≥1, the mortality rate was observed to be very low in the present study.

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AB0662 SUBGLOTTIC STENOSIS IN GRANULOMATOSIS WITH POLIANGIITIS

C.E. Pena¹, F. Giordano², L. Garcia¹, C. Costi¹, G. Pendon², A. Vulcano¹, D. Pereira¹, M. Garcia¹. ¹Rheumatology, HIGA San Martin La Plata; ²Rheumatology, Hospital Gutierrez La Plata, La Plata, Argentina

Background: The characteristic lesion of laryngotracheal involvement in granulomatosis with polyangiitis (GPA) is subglottic stenosis (SGS), due to active disease or from chronic recurrent inflammation, 8% to 23% during the course of GPA and as initial manifestation in 1% to 6% of cases.

Objectives: Describe clinical features and treatment of patients with SGS

Methods: We retrospectively reviewed the medical records of 46 patients with SGS due to GPA diagnosed at a Rheumatology department (2000–2017).

We retrospectively reviewed the medical records of 46 patients with SGS due to GPA diagnosed at a Rheumatology department (2000–2017).

Results: 6/46 patients with GPA diagnosis presented SGS (13%). 83% women, mean age of 37.8 years (DS ±14.8). Mean time between the presentation of the GPA and the diagnosis of SGS:18 months. Most frequent clinical manifestations of SGS: dyspnea (83%), stridor (83%), dysphonia (50%), cough (33%), bronchospasm (16%). One patient presented SGS as initial manifestation of the disease. Mean BVAS:14. Two patients presented such complication with evidence of systemic manifestations. Re stenosis was observed in one patient.

Treatment: IV CYC 83%, oral CYC 16%, methylprednisolone (MP) 83%, oral steroids, plasma exchange (16%) in each one. Endoscopic intervention and balloon dilatation 33%, and intralesional Mitomycin 16%.

Conclusions: 13% patients with GPA presented SGS, being in the majority of cases associated with other manifestations of the disease. However, 67% had no signs of systemic involvement (localised disease). Local treatment was necessary in only 33% because there was a good response to IS in 67% of the cases.

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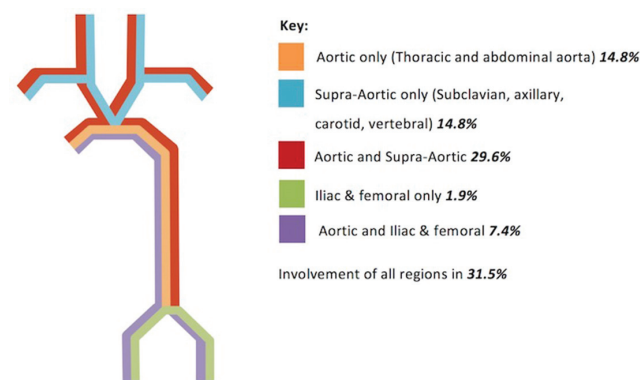
AB0663 18F-FDG-PET/CT DISEASE DISTRIBUTION IN A LARGE VESSEL VASCULITIS COHORT – SUPPORTS VASCULAR ULTRASOUND AS A SCREENING AND DIAGNOSTIC TOOL

F. Coath¹, F. Laskou¹, S. Moghu², A. Sidhu¹, S. Banerjee¹, T. Aung¹, S. Jain², J. Mo², B. Dasgupta¹. ¹Rheumatology; ²Radiology, Southend University Hospital, Westcliff-On-Sea, UK

Background: Subsets of GCA have extensive vascular involvement, termed Large Vessel GCA (LV-GCA), seen in 12%–37% depending on imaging used. These patients have higher relapse rates and are more often refractory to glucocorticoids (GC), requiring steroid-sparing treatment to minimise GC toxicity and vascular complications. Diagnosis is reliant upon imaging, given the relative inaccessibility of tissue for histological analysis beyond temporal artery biopsy (TAB). If axillary and subclavian arteries are often affected, vascular ultrasound could be an effective screening tool.

Objectives: To determine disease distribution in the Southend Large Vessel Vasculitis (LVV) cohort, particularly levels of axillary and subclavian involvement.

Methods: The cohort has sixty-five patients (2010–2017), from which sixty ¹⁸F-FDG-PET/CT scans were performed. They were retrospectively examined by two nuclear radiologists. Vascular involvement was determined by consensus opinion at six territories (thoracic-aorta, abdominal-aorta, subclavian, axillary, carotid, vertebral, and iliac and femoral). Six negative scans were excluded from analysis, with LVV confirmed from other evidence (ultrasound, computed tomography). Of these, four were on concurrent GC (7–12 mg prednisolone), raising the possibility of “false negatives”. Nineteen scans were positive despite GC (1–60 mg prednisolone). Higher doses tended to be of short duration, being unable to postpone due to clinical necessity, but exceeded the 3 day limit at which FDG signal starts to attenuate.



Abstract AB0663 – Figure 1. Disease distribution via ¹⁸F-FDG-PET/CT

Results: 14.8% (n=8) had isolated supra-aortic disease, 14.8% (n=8) had isolated aortic disease, and 29.6% (n=16) had involvement in both (Fig 1). 72.2% of