higher prevalence of mononeuritis multiplex or polyneuropathy
valence of relapses (P.L. Paggiaro2, S. Sellari Franceschini3, S. Bombardieri1

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Introduction

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ESR, CRP, IgE and eosinophil count could not discriminate between
times of remission vs. active disease. Eotaxin-3 levels and eosinophil
counts were, however, significantly higher in patients not using
glucocorticoids during the 28 days before study visit, regardless of
disease activity. Serum eotaxin-3, but not the other biomarkers, was
significantly elevated during active disease in patients not using
glucocorticoids but not elevated in active disease when the patient
was taking glucocorticoids.

Discussion.— All tested biomarkers failed to clearly differentiate active
and inactive disease. Eotaxin-3 may be useful in relapsing EGPA patients
not receiving glucocorticoids before serum sampling.

Conclusion.— Defining biomarkers of relapses in patients with EGPA
remain a challenge especially during times of glucocorticoid use.

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P44 Eosinophilic granulomatosis with polyangiitis (EGPA): Clinical and immunologic expression in a single center cohort
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Introduction.— The purpose of this study was to determine whether
epidemiologic, clinical, and analytical features might modulate disease
expression in a single center cohort of patients with eosinophilic
granulomatosis with polyangiitis (EGPA).

Methods.— Patients with a diagnosis of EGPA made according to the
1990 ACR classification criteria for the disease were enrolled in this
retrospective study. Data collected included:
– gender;
– age at diagnosis;
– cumulative clinical features retrospectively assessed according to the
Birmingham Vasculitis Activity Score (BVAS) glossary.
The following parameters were also recorded: eosinophil count, ANCA
status, rheumatoid factor (RF) positivity. Statistical analysis was per-
formed using SPSS 13 (SPSS Inc., Chicago IL, USA). A 2-tailed value of
P < 0.05 was taken to indicate statistical significance.

Results.— Forty-seven EGPA patients (23F; mean age 47 ± 15 yrs; mean
follow-up 7 ± 5 yrs) were enrolled. The prevalence of the clinical and
laboratory features observed in our EGPA cohort was consistent with the
larger cohorts of the literature. More specifically, ANCA-MPO were
formed in a single center cohort (EGPA): Clinical and immunologic expression
in a single center cohort

Forty-seven EGPA pts (20F; mean age 57.7 ± 14 yrs) were
enrolled in this cross-sectional study. Fiber-optic nasal endoscopy was
performed in all cases leading to the following diagnosis:
– normal;
– allergic (AR) or non allergic rhinitis (NAR);
– CRSwNP or CRSsNP. In all cases, nasal cytologic analysis was per-
formed. NAR, CRSwNP and CRSsNP pts were further subclassified accordingly to
predominant nasal cellular population. The impact of sinonasal involve-
ment on QoL was evaluated by the SF-36 and the Sino-Nasal Outcome
Test-22. Correlations between the different variables were analyzed
using linear regression and the Spearman coefficient (P < 0.05).

Results.— AR was diagnosed in five, CRSsNP in nine, CRSwNP in 13, NAR
in eight patients [of which five with eosinophils (NARES), two with
neutrophils (NARNE), and one without any cytological alteration], and
normal in only two patients. Health-related QoL was deeply impacted by
sinonasal involvement (mean SNOT22: 26.9; mean ISF-36: 42.2;
mean ISM-36: 49.8).

Conclusion.— CRS represents the “clinical prototype” of ENT involve-
ment in EGPA. Sinonasal involvementgreatly affects patients’ QoL, therefore,
multidisciplinary efforts are required in order to optimize nasal symp-
toms treatment and to improve the management of EGPA patients in
clinical practice.

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P45 Sinonasal involvement in patients with eosinophilic granulomatosis with polyangiitis (EGPA, ex Churg Strauss Syndrome): A modern look to an ancient problem
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Introduction.— “Paranasal sinus abnormalities” are commonly reported
in granulomatosis with polyangiitis (EGPA) representing one of the six
1990 ARA classification criteria for the disease. Although frequently
encountered, the definition of “paranasal sinus abnormalities” in EGPA
has been based on a number of often-vague patients’ symptoms
making it uncertain to assess the real prevalence, the presentation
pattern and the clinical course of ENT involvement in EGPA. The purpose
of this study was to describe the frequency and the clinical presentation
of ENT involvement in a series EGPA patients. We focused on sinonasal
involvement and on cytological analysis, as a tool to better-diagnosed
sinonasal inflammatory diseases.

Methods.— Thirty-seven EGPA pts (20F; mean age 57.7 ± 14 yrs) were
enrolled in this cross-sectional study. Fiber-optic nasal endoscopy was
performed in all cases leading to the following diagnosis:

P46 A case of eosinophilic granulomatosis with polyangiitis (EGPA) who relapsed with saddle nose
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Introduction.— Saddle nose is a typical symptom of Granulomatosis with
polyangiitis (GPA). We report here an interesting case of EGPA, who was
treated with saddle nose deformity as an expression of relapse.
induced remission once and relapsed with saddle nose. We also discuss the pitfalls of Watts' algorithm for classification of ANCA-associated vasculitis.

**Methods.** A 73-years-old female had a history of asthma. In October 2009, she suffered fever, abnormal sensation in both legs, and leg edema. She had 2,4600 μl of peripheral eosinophils and was seropositive for MPO-ANCA. They diagnosed her with EGPA, and she started to be treated with prednisolone and transferred to our hospital. On admission, she showed paresthesia, leg edema, marked eosinophilia, renal dysfunction, proteinuria and hematuria, reduced nerve conduction velocity. However, remission induction was achieved after several months.

When the dose of PSL was tapered, she suffered a relapse with nasal bleeding, saddle nose, and elevation of ANCA titer. However, there was no eosinophilia at the relapse. We treated her with prednisolone and intravenous cyclophosphamide for the second remission.

**Results.** Saddle nose is associated with GPA. CT scan of paranasal cavity suggested that her sinusitis was due to GPA. It is very rare for an EGPA patient to show saddle nose. There has been only one report in which saddle nose was seen in a patient of EGPA (Takizawa et al., 1989). Their case was very similar to our case, even though ANCA could not be measured in those days.

**Discussion.** Using Watts' algorithm, we diagnose first with EGPA and then check surrogate markers for GPA. Thus, if a patient fulfills the criteria of EGPA, we diagnose with EGPA even though they have surrogate markers for GPA. In Asian countries, there are much more MPO-ANCA positive GPA compared to Western countries. They tend to be diagnosed with EGPA, because GPA patients sometimes have a history of asthma and mild eosinophilia.

**Conclusion.** We experienced an interesting case of EGPA who relapsed with the symptom of saddle nose.

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**Epidemiology**

P47

**Referral causes and initial diagnosis of ANCA-associated vasculitides in a pulmonary tertiary centre. Retrospective study in 90 patients**

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**Introduction.** No studies have addressed referral causes and initial diagnosis of ANCA-associated vasculitides (AASV) in a respiratory centre. We have studied this issue and present the main clinical characteristics of 90 patients with AASV.

**Methods.** Retrospective, descriptive review of patients with final diagnosis of AASV based on the ACR criteria and the 1994 Chapel Hill Consensus Conference Nomenclature from 1982 to 2010.

**Results.** Ninety patients (74 GPA, ten MPA, six CSS) were studied. Only one had an initial suspicion of an AASV. Main categories considered as first diagnosis were infectious (n=50), rheumatological diseases (n=19), neoplastic (n=9, only for GPA patients) and other in 12. Mean time elapsed from initial suspected to definitive diagnosis was 30 months. When we compared the clinical manifestations observed in our GPA patients with a similar series derived from a respiratory center (n = 77) [1], we found that SGS was significantly more common in our study (31% vs 2.5%), while cough (40 vs 78%), rhinitis (20% vs 42%), hemoptysis (18% vs 39%) and chest pain (3% vs 32%) were less frequent. After a mean follow-up of 22 months, 83% of patients were alive with remission being achieved in 87% and response in 9%. Seven patients died, mostly from infectious complications.

**Discussion.** The majority of descriptions regarding respiratory disease in AASV are in the context of data from cohorts attending nephrological or rheumatological units. Our study has addressed the initial diagnosis considered in patients with an AASV in a respiratory centre in where the correct diagnosis was not apparent for any of the cases and alternative diseases, mostly of infectious origin, were considered first.

**Conclusion.** On referral or arrival to a respiratory centre, our patients were thought to have other diseases. This resulted in delay until correct diagnosis was reached. By making our data available, we aim to revert this condition by expanding the knowledge among our respiratory specialists on the modes of presentation of AASV patients.

**Reference**


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P48

**ANCA in Auckland 1998–2008**

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**Introduction.** ANCA associated systemic vasculitis (AASV) is a rare condition. Here we describe in detail the clinical features and outcomes of patients with AASV from Auckland, New Zealand.

**Methods.** All positive results for ANCA antibody between 01/01/1998 and 01/01/2008, were determined by electronic search. A retrospective case notes review was undertaken. Data was censored at 01/01/2010.

**Results.** One hundred and twenty-four patients were identified. 66 Females. Mean age at presentation was 58.8 yrs (range 9.3–94.2 yrs). One hundred were European, seven NZ Maori, five Pacific Peoples, 12 of other ethnicity. Using Pearson’s chi squared test, it was less likely for NZ Maori and Pacific People to have AASV than Europeans (P < 0.002). A chi-square goodness of fit test showed no excess by month of presentation or season.

Using the EMEA classification [1], 70 cases were GPA, 42 MPA and 12 CSS.

Two patients had a pANCA with no ELISA available. Using ELISA there were 59 cases of MPO+ve ANCA and 63 of PR3+ve ANCA.

At presentation mean serum creatinine was 204 μmol/L (range 50–1150 μmol/L); median creatinine 106 μmol/L. Seventy-seven patients had haematuria and 64 had proteinuria.

CXR was performed in 123 patients at presentation: 43 had abnormalities attributable to AASV, 13 had alveolar haemorrhage; ten had respiratory failure.

One hundred and sixteen patients received corticosteroids, 86 cyclophosphamides, ten plasma exchanges.

Thirty-four patients developed cancer, 12 cardiovascular diseases, 16 cerebrovascular events, four PVD.

Fifteen patients developed end stage kidney disease requiring long-term renal replacement therapy.

Fifteen were lost to follow-up. Two patients died acutely of respiratory failure. 47 died in total.

**Discussion.** For a New Zealand cohort, we found a similar male:female ratio with an older age group.

We saw equal number of MPO and PR3 cases with GPA the predominant syndrome.