**P61**

**Successful pregnancy in patients with granulomatosis with polyangiitis (Wegener’s) after rituximab treatment**

P. Novikov, E. Kuznetsova, O. Borodin, A. Meshkov, S. Moiseev
First Moscow State Medical University, Moscow, Russia

**Introduction.**– GPA frequently develops in the elderly patients though it can manifest at younger age. High activity of disease and immunosuppressive treatment increase the risk of unfavourable outcomes of pregnancy. RTX is highly effective in GPA patients but its effects on reproductive function are not well studied.

**Patients.**– We present the case of successful pregnancy in GPA patient treated with RTX.

**Results.**– In 26 years old female GPA manifested with bilateral acute otitis and lung infiltrates. Antibiotics were ineffective. Later she developed ulcerative rhinitis and fever. Laboratory tests showed the presence of acute inflammation, urinary changes (red blood cells, protein, casts) and PR3 ANCA. Nasal biopsy revealed granulomatosis and vasculitis. The remission of GPA was achieved after treatment with oral PRED and intravenous CYC (every 3 weeks for 6 months). The patient continued treatment with low-dose PRED and AZA but after 10 months she developed the relapse of GPA with upper and lower respiratory tract involvement. RTX administration (four infusions of 500 mg with weekly intervals) rapidly induced the remission of GPA. All drugs except PRED 5 mg daily were discontinued because the patient planned pregnancy. She developed pregnancy within 8 months after RTX administration. The course of gestation was uneventful. PR3 ANCA level increased at 6 months. She delivered healthy baby with normal B-cell counts in term (Apagar 9/9). Immediately after delivery CT scan showed no changes in the lungs and nasal cavity but within 4 months the patient developed the relapse of ulcerative rhinitis and was effectively retreated with RTX.

**Conclusion.**– According to the current EULAR guidelines the indications for RTX administration in GPA patients include inefficacy or intolerance of standard treatment or frequent exacerbations. RTX should be preferably in younger patients who consider pregnancy. Our case shows the possibility of successful pregnancy in young female with GPA within few months after RTX administration.

http://dx.doi.org/10.1016/j.lpm.2013.02.132

**P62**

**Distinct eicosanoid profile in exhaled breath condensates from granulomatosis with polyangiitis (Wegener’s) patients**

W. Szczeklik, M. Sanak, J. Sznajd, B. Jakiela, B. Sokolowska, M. Kaszuba, A. Sawina, J. Musial
Jagiellonian University Medical College, Krakow, Poland

**Introduction.**– Granulomatosis with polyangiitis (WGA, Wegener’s) is a systemic vasculitis affecting mostly small vessels, typically involving the upper respiratory tract, lungs and kidneys. The hallmark of the respiratory tract pathology is granulomatous inflammation, and eicosanoids play an important role in it. We aimed to evaluate the eicosanoid profile in GPA patients, using a noninvasive method – exhaled breath condensate (EBC) analysis.

**Patients.**– EBC from 27 GPA patients and 30 healthy controls (HC) were assessed quantitatively for 20 eicosanoids by mass spectrometry by a high-performance liquid chromatography–tandem mass spectrometry (HPLC-MS/MS).

**Results.**– In the GPA group LTB4, PGD2, and 11-dehydro-TXB2 were elevated when compared to HC, whereas: 5-HETE, LTC4, trans LTC4, eosins (C4, D4, E4), 9α, 11β-PGF2α and tetranor-PGE-M were decreased (figure S1).

**Conclusion.**– Significant differences in the EBC eicosanoid profiles between the GPA and HC groups were observed. This finding may help us to elucidate the pathogenesis of the respiratory tract inflammation in GPA patients.

**Supplementary data associated with this article can be found on the website of La Presse Médicale** (http://www.em-consule.com/revue/lpm).

Figure 1. Differences between eicosanoid concentrations in exhaled breath condensate from patients with granulomatosis with polyangiitis (GPA) and healthy controls (HC). Abbreviations: M: metabolite; **: < 0.01.

http://dx.doi.org/10.1016/j.lpm.2013.02.133

**P63**

**Alveolar hemorrhage, rectus muscle hematoma, and gastrointestinal bleeding accompanied by crescentic glomerulonephritis in a patient with PR3-ANCA disease**

Y. Cao
China-Japan Friendship Hospital, Beijing, China

**Introduction.**– This is the first report of a case of GPA accompanied by alveolar hemorrhage, rectus muscle hematoma, and gastrointestinal bleeding.

**Patients.**– A 67-year-old female was admitted to our hospital because of general fatigue, fever, and edema for one month, and hemoptysis, dyspnea for 1 week. Scr was rapidly increased to 4.0 mg/dL with hematuria and proteinuria. CT revealed large areas of ground glass opacity in the right lung; hence the hemoptysis was due to alveolar hemorrhage. Sputum culture revealed the presence of Staphylococcus. The titer of PR3-ANCA was 340 U/L, and he was diagnosed with ANCA disease (BVAS 48) associated with pneumonias. Thus, she was treated with antibiotics and plasma exchange (PE). However, after the second PE, she suddenly complained of abdominal pain and swelling in the right rectus muscle region with hypotension. Her Hb dropped from 8.8 to 4.5 g/dL in the subsequent hours. Abdominal CT showed an irregular low-density mass in the right muscle, so he was diagnosed as rectus muscle hematoma. Her condition was improved after receiving a transfusion and supportive therapy. Then, she was started on methyl-prednisolone pulse therapy followed by the oral prednisolone and cyclophosphamide resulted in improvement of hemoptysis with gradually decreased Scr. However, she developed tinnitus and bloody nasal discharge and diagnosed as frontal sinuses by CT. Therefore, we gave her the third PE. Unfortunately, she developed tarry stools and her Hb dropped soon. Endoscopy showed two duodenal ulcers, and the biopsy specimen revealed angitis of the duodenum. Another methyl-prednisolone pulse therapy, inhibition of gastric acid and transfusion were given. Renal biopsy showed the presence of crescent-shaped...
Characterization of F-18 fluorodeoxyglucose PET/CT in granulomatosis with polyangiitis

D. Nelson, G. Johnson, R. Cartin-Ceba, U. Specks
Mayo Clinic, Rochester, USA

Introduction.– Granulomatosis with polyangiitis (GPA) is a rare autoimmune disease that is characterized by granulomatous inflammation and small vessel vasculitis that primarily involves the upper airways, lungs, and the kidneys. Early and accurate diagnosis and assessment of the extent of the disease is important for treatment decisions. F-18 fluorodeoxyglucose (FDG) positron emission tomography (PET) is an imaging technique that is increasingly being used in evaluation and management of other types of vasculitis (giant-cell arteritis). The role and characterization of FDG-PET/CT has not been reported for GPA.

Patients.– We identified 10 patients with GPA who underwent FDG-PET/CT scanning over a 7-year period from January 2005 to December 2012. In all 10 patients the FDG-PET/CT was performed for suspected or known malignancy. The presenting clinicoradiologic features including FDG-PET/CT scan and chest CT scan findings were analyzed.

Results.– Differentiation between inflammatory and malignant lesions was not able to be determined based on FDG-PET/CT imaging. Max SUV was compared in patients who had malignant lesions and those with inflammatory lesions secondary to GPA and no significant difference was noted. GPA lesions of the respiratory tract and lung were more clearly demonstrated than other lesions. FDG uptake noted within vessels that was not previously appreciated. Eight of 10 patients had uptake noted in the lung, and 4 out of 10 had uptake in the sinuses. Three patients had FDG uptake noted within vessels that was not previously appreciated. In seven out of 10 patients the PET/CT guided the location of the biopsy and led to diagnosis. Two patients had follow up FDG-PET/CT, which demonstrated decreased FDG uptake after treatment.

Conclusion.– FDG-PET/CT cannot differentiate between malignant and inflammatory lesions in patients with GPA. However, it is a feasible modality to evaluate GPA lesion activity, identify new areas of involvement and help to guide biopsy location.

Further readings
http://dx.doi.org/10.1016/j.lpm.2013.02.135

P65 Presentation and management of granulomatosis with polyangiitis (Wegener’s) (GPA) central nervous system (CNS) involvement

G. De Luna, B. Terrier, P. Charles, C. Pagnoux, X. Puechal, P. Cohen, L. Mouthon, L. Guillemin
Cochin, Paris, France

Introduction.– GPA, a small-sized–vessel vasculitis, commonly involves ear, nose & throat (ENT), lungs and kidneys, and rarely, the CNS. The presentation, management and outcome of GPA CNS involvement were evaluated.

Patients.– We retrospectively reviewed the charts of 16 patients (12 men) with: GPA satisfying ACR and/or Chapel Hill criteria; and, after excluding other causes, GPA CNS involvement manifesting as pachymeningitis, meningitis, stroke, spinal cord involvement or hypophysial involvement.

Results.– Mean respective ages at GPA diagnosis and onset of CNS involvement were 43 and 47 years. The latter was present in nine (56%) patients at GPA diagnosis, and appeared in the seven others after a median follow-up of 24 months. Headache was the main symptom (67%), with motor and sensory impairments noted in 33 and 27% respectively. CNS involvements were: pachymeningitis (n = 8: seven cranial and one spinal cord), ischemic (n = 4) or hemorrhagic stroke (n = 2), cerebral vasculitis (n = 2), and/or hypophysial involvement (n = 2). Extra-CNS manifestations included ENT (75%), lungs (60%), peripheral nerve(s) (40%) and kidneys (33%). ANCA detected in 12/16 (75%) patients had PR3 (n = 7) or MPO (n = 5) specificity.

Induction therapy comprised corticosteroids (CS, 100%) and IV (69%) or oral CYC (43%) or rituximab (43% each). Maintenance therapy consisted of CS (100%) and azathioprine (63%), or methotrexate or rituximab (13% each).

CNS involvement responded clinically in 12/16 (75%). Relapsing and/or refractory CNS GPA in seven patients was treated with IV CYC or rituximab (43% each) or oral CYC (14%). No patient died during follow-up, but 64% had persistent neurological sequelae.

Conclusion.– Our series highlights the heterogeneity of CNS involvement in GPA. Despite initial severe disease, conventional therapy obtained clinical improvement in 75% of the patients. Rituximab should be evaluated for refractory and/or relapsing CNS GPA.

http://dx.doi.org/10.1016/j.lpm.2013.02.136

P66 Pituitary dysfunction in granulomatosis with polyangiitis

R. Cartin-Ceba, E. Singh, T. Peikert, K. Keogh, D. Erickson, U. Specks
Mayo Clinic, Rochester, USA

Introduction.– Pituitary gland involvement has been rarely reported in granulomatosis with polyangiitis (GPA).

Patients.– Retrospective case-series describing clinical features, pituitary imaging, hormonal evaluation, and treatment of all patients with pituitary involvement by GPA evaluated at Mayo Clinic, between 01/1996 and 12/2011.

Results.– Pituitary involvement was noted in nine patients (five women). Median age at diagnosis of both GPA and pituitary involvement was 48 (range 28–68). All patients were c-ANCA/PR3 positive. Pituitary involvement frequently represented an incidental imaging finding during the evaluation of headache or sinusitis (six patients). Three