hemostatic effect. For uncontrolled hypoxia, he was transferred to the ICU and intubated.

Pulmonary bleeding stopped after rFVIIa administration iv. Bronchoscopy on day 8 was negative for bleeding. He was depending on mechanical ventilation but the arterial oxygen pressure (pO2)/(FiO2) ratio increased the following day. Over the next 10 days, the situation improved and pt was discharged from the ICU.

No thromboembolic complications were observed after administration of rFVIIa iv.

Conclusion.— This case suggests that in levamisole-adultered cocaine induced syndrome, severe lung and renal involvement can occur and rFVIIa iv. should be considered in severe DAH not responding to therapy.

http://dx.doi.org/10.1016/j.lpm.2013.02.050

A49
ANCA-associated vasculitis (AAV) and thrombotic microangiopathies (TMA): A retrospective histological study

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Introduction.— A number of case reports of vasculitis with associated TMA are progressively reported in the literature. Moreover studies in animal models suggest that complement activation is crucial in the pathogenesis of ANCA-associated vasculitis (AAV) [1], supplying a possible predisposing pabulum for TMA appearance. At our knowledge no data are available describing the possible association between vasculitis and TMA. We studied retrospectively our historical AAV cases to document the presence of histological signs of TMA.

Patients.— We retrospectively examined a series of 39 consecutive patients diagnosed with renal biopsy as having AAV between 1990 and 2012. Patients had a mean follow-up of 86 months. Renal histology was reviewed by an independent pathologists, blinded for renal outcome. Results.— Ten out of thirty-nine patients died during follow-up. Six out of thirty-nine (15%) patients were identified as having clear signs of TMA. Four out of six patients of the TMA group had often an associated histological evidence of fibrinoid necrosis while the non TMA group presented an overall greater risk for renal failure on the basis of recently risk stratification scores in renal vasculitis; nonetheless the TMA group showed a significantly worse renal outcome ($P < 0.05$) (figure 1).

Discussion.— Our preliminary data suggests that, similarly to other glomerulonephritis, TMA should be evaluated as an associated histological pattern in renal vasculitis. TMA seems to determine a worse renal prognosis; consequently recognition of TMA associated features may allow a more appropriate management (i.e plasmapheresis) of acute renal failure in these cases.

Reference

http://dx.doi.org/10.1016/j.lpm.2013.02.051

A50
The relative risk of aortic aneurysm in patients with giant cell arteritis compared with the general population of the UK

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Introduction.— Giant cell arteritis (GCA) is the most common systemic vasculitis; small studies have demonstrated 22% of these patients will develop an aortic aneurysm [1]. The British Society of Rheumatology and the European League Against Rheumatism has highlighted the need for a precise estimate of the increased risk of aortic aneurysm to inform definitive advice regarding screening of patients with GCA [2,3].

Patients.— This parallel cohort study includes 6999 patients with GCA from the UK General Practice Research Database (GPRD). Non-GCA patients were matched on a 6:1 ratio on: GP practice, year of birth and gender. A competing risk model using aortic aneurysm as the primary outcome and death as the competing risk was used to determine the relative risk (subhazard ratio) between non-GCA and GCA subjects, after adjustment for cardiovascular risk factors.

Results.— Comparing the GCA cohort with the non-GCA cohort; after adjustment for BMI, smoking, alcohol, hyperlipidaemia, lipid lowering medication, hypertension, anti-hypertensives, diabetes, cardiovascular disease, cerebrovascular disease and peripheral vascular disease; the subhazard ratio for aortic aneurysm (95% CI) was 1.92 (1.52 to 2.41). Significant predictors of aortic aneurysm were as follows: being a smoker 3.37 (2.61 to 4.37), previous history of diabetes 0.32 (0.19 to 0.56) and previous history of cardiovascular disease 1.98 (1.50 to 2.63). In a multivariable model of the GCA cohort alone, male gender 2.10 (1.38 to 3.19), smoking 3.79 (2.20 to 6.53), and diabetes 0.19 (0.05 to 0.77) were significant predictors of aortic aneurysm.

Discussion.— This is the largest cohort study of patients with GCA to date, and demonstrates a two-fold increased risk of aortic aneurysm. Other risk factors for aortic aneurysm, including male gender, age, and smoking, and a protective effect of diabetes, are however also important in GCA patients.

Conclusion.— GCA should be considered within the context of other risk factors for aortic aneurysm, rather than needing separate screening.
Risk of cardiovascular disease early and late after a diagnosis of giant cell arteritis

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Introduction.—Despite well-documented involvement of large vessels in giant cell arteritis (GCA), the risk of clinically important cardiovascular events is not known. The objective was to evaluate the risk of incident myocardial infarction (MI), cerebrovascular accidents (CVA), and peripheral vascular disease (PVD) in individuals with incident GCA in a general population context.

Methods.—We conducted a cohort study using a UK primary care database containing records from 1986 to 2011. GCA diagnoses, outcomes, and cardiovascular risk factors were identified from electronic medical records. We conducted separate sub-cohort analyses for:
- MI;
- CVA;
- PVD, excluding individuals with prevalent disease at baseline for each analysis.

We estimated hazard ratios (HRs) comparing GCA with age-, sex-, and entry time-matched comparison cohorts, adjusting for potential cardiovascular risk factors.

Results.—Among 3533 individuals with GCA (73% female, mean age 73 years), the incidence rates (IRs) of MI, CVA, and PVD were 10.0, 8.0 and 4.2 per 1000 person-years (PY), versus 4.9, 6.3 and 2.0 per 1000 PY in the comparison cohort. The corresponding adjusted HRs were 1.97 (95% CI 1.42 to 2.75) for MI, 1.77 (95% CI 1.24 to 2.53) for CVA and 2.76 (95% CI 1.75 to 4.36) for PVD. These HRs were more pronounced in the first month after the diagnosis of GCA: MI (HR = 11.89, 95% CI: 2.40–59.00), CVA (HR = 3.93, 95% CI: 1.76–8.79), and PVD (HR = 3.86, 95% CI: 0.78–19.17).

Conclusion.—These findings provide the first general population-based evidence that GCA is associated with an increased risk of developing MI, CVA, and PVD. Further research into the causes of this increased risk in GCA, including how current medications used in GCA may alter cardiovascular risk, would be valuable.

http://dx.doi.org/10.1016/j.lpm.2013.02.053

A52

Large vessel vasculitis: Estimating disease activity in patients with thoracic aortic aneurysms

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Introduction.—Thoracic aortic aneurysms may be a sign of ongoing inflammation or the sequelae of previous damage to the arterial wall. Ongoing inflammation in the aorta is usually estimated indirectly using clinical features, inflammatory markers and imaging characteristics. Whether these surrogates accurately reflect inflammation within the vessel wall is not known.

Methods.—We retrospectively reviewed the clinical, laboratory and imaging data for patients in whom aortitis was histologically demonstrated at the time of reconstruction for thoracic aortic aneurysms (May 2011–December 2012). ACR classification criteria were used to define vasculitis subtype. Pre- and postoperative assessments of disease activity were estimated over time using predefined criteria.

Results.—Among 126 consenting study patients who underwent repair of thoracic aortic aneurysms, 14 (11%) had histological features of aortitis. Thirteen specimens had active inflammatory infiltrates, and one had features of healed aortitis. Seven patients met criteria for Giant Cell Arteritis, two for Takayasu’s arteritis, and five for focal isolated aortitis. Median duration of disease prior to surgery was 35.5 months (range 0–180). At the time of surgery, median ESR and CRP were 11 mm/hr (range 2–38) and 1.0 mg/dL (range 0.2–1.7), respectively, and angiograms revealed new lesions in 3/14 patients. Nine patients (64%) were judged to be in clinical remission, in three the degree of disease activity was uncertain, and two met criteria for active disease. Seven patients were treated with prednisone within 12 weeks of surgery [median dose = 20 mg/d (range 2–55 mg)].

Discussion.—About 2/3 of patients found to have aortitis do not have clinical, laboratory or imaging indications of active disease at the time of surgery.

Conclusion.—Indirect estimates of thoracic aortic inflammation are insensitive in patients with aneurysms.

Further reading

http://dx.doi.org/10.1016/j.lpm.2013.02.054

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Assessment of disease activity in large vessel vasculitis: Initial results of an international Delphi exercise

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