Discussion.— We found IgAV present in one third of all DIF+ LCV cases, often in the context of an infection. The prognosis is favourable in most cases.

Conclusion.— The high frequency of bacterial infections seen before disease onset in this cohort support that further investigation may be warranted into whether a link exists between infection and adult IgAV.

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P143
Macrophage activation syndrome in vasculitis: An underrecognised complication?

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Introduction.— Macrophage activation syndrome (MAS) is potentially fatal in rheumatic diseases. We retrospectively studied the clinical picture, treatment and outcome of MAS in vasculitis patients seen in a referral rheumatology unit in Transylvania, Romania.

Methods.— The charts of vasculitis patients admitted or seen ambulatory between 2000 and 2012 were reviewed. MAS was defined according to Ravelli preliminary criteria for juvenile idiopathic arthritis.

Results.— We identified four MAS cases, three in previously diagnosed vasculitis (microscopic polyangiitis with polychondritis, granulomatosis with polyangiitis, undifferentiated vasculitis) and one inaugural of Behçet’s disease. In all cases, high fever, lethargy, pancytopenia, elevated transaminases and hyposodemia were noted. Ferritin was elevated in the three cases available. Hepatosplenomegaly and/or adenomegaly were seen in all cases. Bone marrow showed hemophagocytosis in two of three cases performed. Coagulation abnormalities were noted in three patients, along with antiphospholipid syndrome and transverse myelitis in the granulomatosis case. The trigger was infectious in two patients (pneumonia with S. aureus and E. coli). Methylprednisolone pulse therapy, cyclosporin, broad spectrum antibiotics, life support, transfusions, anticoagulation were employed, successful in two cases.

Discussion.— MAS can be inaugural in vasculitis and may mimic a drug-induced complication or a hematologic malignancy. Therapy cessation, surgery and infections are precipitating factors. Low ESR can be misleading.

Conclusion.— MAS is rare, but life-threatening in vasculitis. A high index of suspicion and prompt therapy are necessary for outcome improvement.

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P144
Retroperitoneal fibrosis at the onset of ANCA associated vasculitis: The risk of a delay in the diagnosis of vasculitis

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Introduction.— Retroperitoneal fibrosis (RPF) is characterised by the development of fibroserotic tissue in the retroperitoneum which leads to encasement of ureters and blood vessels. It is usually “idiopathic”, rarely it’s due to drugs, malignancies, infections, or autoimmune diseases including ANCA associated vasculitis (AAV).

Results.— We describe three cases of AAV with RPF as initial manifestation misinterpreted as idiopathic.

Three male patients (Pt1 42 yo; Pt2 49 yo, Pt3 59 yo) were admitted with back pain, deep vein thromboses (DVT) and monolateral hydronephrosis (Pt1); with DVT, bilateral hydronephrosis and acute renal failure (ARF) (Pt2); monolateral hydronephrosis and ARF (Pt3). In Pt1 and 2, abdominal CT scan and MRI revealed homogeneous dense fibrous tissue around the aorta, with inferior vena cava and iliac veins compression and thrombosis and monolateral/bilateral ureteral involvement. In Pt3 renal US revealed monolateral hydronephrosis, he was submitted to ureteral stenting, no further evaluations were planned.

On the basis of diagnosis of idiopathic RPF, all pts started oral glucocorticoid therapy at mean doses of 0.5 mg/kg/day with symptomatic relief; in Pt2 obstructive ARF resolved. After 2 and 6 months respectively, Pts1 and 2 developed AAV overt manifestations. Pt 1: RPGN, multineuritis and other vasculitis related signs) can be hidden by RPF manifestations and by steroids therapy given to treat “idiopatic RPF”. In Pts1, ARF didn’t improve and he started chronic hemodialysis. He was maintained on low dose steroids therapy for long time. Seven years later, while still on chronic dialysis, he developed persistent fever, malaise, arthralgias. CT scan and MRI revealed a RPF with DVT.

Conclusion.— RPF can be the initial manifestation of AAV. Clinical manifestation of AAV (RPGN and other vasculitis related signs) can be hidden by RPF manifestations and by steroids therapy given to treat “idiopathic RPF”. RPF at the onset of AAV entails the risk of a delay in AAV diagnosis and treatment.

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