I. Felea³, I. Filipescu⁷, S. Rednic⁷, A. Bojan⁸

Introduction

8. Iuliu Hatieganu University of Medicine and Pharmacy Cluj,
7. Iuliu Hatieganu University of Medicine and Pharmacy Cluj,
6. Emergency County Teaching Hospital Cluj, Immunology Dept,
5. Iuliu Hatieganu University of Medicine and Pharmacy Cluj, 2nd
4. Emergency County Teaching Hospital Cluj, Pathology Dept, Cluj-
3. Emergency County Teaching Hospital Cluj, Cluj-Napoca, Romania
2. Iuliu Hatieganu University of Medicine and Pharmacy Cluj,
L. Damian¹, C. Cismaru², L. Ghib³, M. Muresan⁴, B. Stancu⁵, M. Sfichi⁶,
An underrecognised complication?

Discussion

http://dx.doi.org/10.1016/j.lpm.2013.02.213

P143

Macrophage activation syndrome in vasculitis: An underrecognised complication?

L. Damian¹, C. Cismaru², L. Ghib³, M. Muresan⁴, B. Stancu⁵, M. Sfichi⁶,
I. Felea³, I. Filipescu⁷, S. Rednic⁷, A. Bojan⁸
1. Emergency County Teaching Hospital Cluj, Rheumatology Dept, Cluj-Napoca, Romania
2. Iuliu Hatieganu University of Medicine and Pharmacy Cluj, Infectious Diseases Dept, Cluj-Napoca, Romania
3. Emergency County Teaching Hospital Cluj, Cluj-Napoca, Romania
4. Emergency County Teaching Hospital Cluj, Pathology Dept, Cluj-Napoca, Romania
5. Iuliu Hatieganu University of Medicine and Pharmacy Cluj, 2nd Surgery Dept, Cluj-Napoca, Romania
6. Emergency County Teaching Hospital Cluj, Immunology Dept, Cluj-Napoca, Romania
7. Iuliu Hatieganu University of Medicine and Pharmacy Cluj, Rheumatology Dept, Cluj-Napoca, Romania
8. Iuliu Hatieganu University of Medicine and Pharmacy Cluj, Hematology Dept, Cluj-Napoca, Romania

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 the 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.

http://dx.doi.org/10.1016/j.lpm.2013.02.214

P144

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

S. Possenti¹, F. Londrino², G. Jeannin¹, C. Salviani³, G. Cancarini³, G.A. Gregorini¹
1. UO di Nefrologia e Dialisi, Spedali Civili di Brescia, Brescia, Italy
2. SC di Nefrologia e Dialisi Ospedale S. Andrea, La Spezia, Italy
3. UO di Nefrologia e Dialisi A, Policlinico Umberto I di Roma, Roma, Italy

Introduction.— Retroperitoneal fibrosis (RPF) is characterised by the development of fibrosclerotic 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 a wide lung consolidation; Pt2 developed RPN, multisite renal, cutaneous purpura and episcleritis. Pt2 died in acute phase because of diffuse lung hemorrhage favoured by chronic anticoagulation prescribed at time of DVT.

In Pt3, 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 RMI revealed a RPF with DVT.

In all three pts, ANCA resulted highly positive with anti PR3 specificity.

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.

http://dx.doi.org/10.1016/j.lpm.2013.02.215