

vasculitis, "2": less than five, "3": five or more involved blood vessels/microscopic field with a x20 objective)

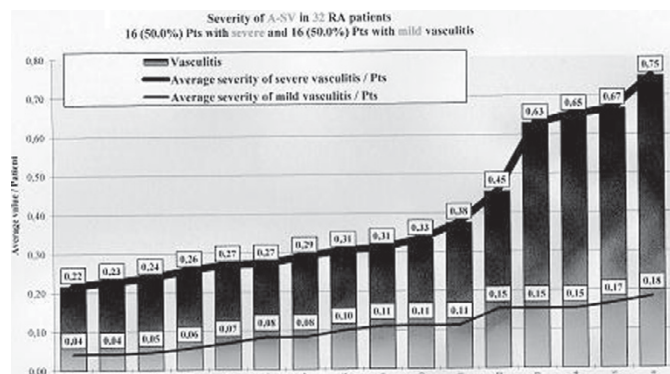
The age at death, and the onset and duration of RA with mild (<0.2/patient), or severe (0.2A-SV, were compared by Student (Welch) t-probe.

Results: 16 (50.0%) of 32 patients had a "mild" degree of A-SV (females 11, avg age of 70.91 years, range 80 – 63, onset of RA: 62.55, avg disease duration: 8.36 years at death; males 5, avg age of 71.6 years, range 83–58, onset of RA: 57.2, avg disease duration: 14.4 years).

16 (50.0%) of 32 patients had "severe A-SV" (females 8, age of 60.88 years, range 82–32, onset of RA: 52.67, avg disease duration: 13.5 years at death; males 8, avg age of 65.0 years, range 78–53, onset of RA: 53.25, avg disease duration: 11.75 years).

Four (12.5%) of 32 patients had "extremely severe A-SV" (with an average cumulative value of severity/RA patient with SV >0.630) (females 1, age of 82.0 years, onset of RA: 62.0, average disease duration: 20.0 years at death; males 3, average age of 69.7 years, range 78 – 59, onset of RA: 60.7, average disease duration: 9.0 years)

Severity of SV in 32 RA patients with A-SV – according to increasing average values of vasculitis/patient – is summarized in Figure.



Conclusions: A-SV is caused by circulating immune complexes in RA. Immune complexes spread via the bloodstream and provoke vasculitis throughout the body.

We found no differences in the linear and basically parallel development of A-SV between patient groups with mild and severe vasculitis.

The progression of vasculitis was the same in both patient groups suggesting differences in production of circulating immune complexes.

Quantitative differences in the production of circulating immune complexes may be related to a "benign" or "aggressive" clinical course of RA, which may be due to genetical and other factors.

In 4 of 32 RA patients the severity of vasculitis showed a "step-wise" growth curve with "extreme severe" A-SV (0.630 or <) according to increasing average values of vasculitis/patient (Figure 1). There was no gradual transition between the "extremely severe" and "severe" degrees A-SV. This profile of "step-wise" general severity in patients with vasculitis may represent a subgroup of patients with a different genetic-immunologic background.

References:

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Disclosure of Interest: None declared

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AB0578 ARTERIAL ANEURYSMS MOROCCAN EXPERIENCE STUDY OF 37 CASES

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Background: Behcet's disease is a systemic vasculitis with a tropism for the venous system. Arterial involvement is uncommon (1%) and mainly represented by aneurysms that can be life-threatening.

Objectives: This retrospective study was conducted in the internal medicine department of the University Hospital Ibn Rochd of Casablanca, over a period of thirty-five years between 1980 and 2016. Where included all the cases of Behcet's disease diagnosed in our service (1618 case).

Methods: We aimed to determine the epidemiological profile, the different possible clinical manifestations and to discuss both prognosis and treatment in such cases.

Results: 37 patients – 32 men and 5 women – presented arterial involvement in type of arterial aneurysm, which represents a rate of 2.35%.

Mean age at diagnosis was 32 years old (ranges 17–54). This complication was the revealing event for Behcet's disease in 2 cases, concomitant in 3 cases and occurring after an average of 6-year-period evolution of the disease in 32 cases.

The aneurysm affected: the pulmonary artery (22 cases), the abdominal aorta (5 cases), the femoral artery (5 cases), the internal carotid artery (2 cases), the iliac artery (2 cases) and the middle cerebral artery (1 case). The aneurysm was associated with venous disease (18 cases), pulmonary embolism (2 cases) and intracardiac thrombus (1 case).

The medical treatment has relying on anticoagulants (6 cases), anti-aggregating agents (9 cases), corticosteroids (36 cases), immunosuppressive drugs – cyclophosphamide (23 cases) and azathioprine (12 cases), while 7 patients underwent surgical intervention.

Evolution was favorable in 23 patients and with negative outcome in 14 patients (9 relapses and 5 deaths).

Conclusions: Arterial aneurysms are the most common arterial complications in the context of Behcet's disease, while the prognosis remains poor in the absence of early and appropriate management (corticosteroids, immunosuppressive agents, surgery).

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AB0579 IMMUNE RELATED DIFFUSE ALVEOLAR HAEMORRHAGE: SINGLE CENTER EXPERIENCE AND LONG TERM OUTCOME – RHEUMATOLOGICAL PERSPECTIVE

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Background: Diffuse alveolar haemorrhage (DAH) is a feature of several immune and nonimmune disorders. Failure to diagnose and treat DAH syndromes in their early stages may lead to acute respiratory failure, CKD and death. Prognosis is poor with in-hospital mortality ranging from 20% to 100%. Immune related DAH is monophasic and if treated early and achieved remission, long term outcome is good.

Objectives: To evaluate the therapeutic response and long term outcome in patients with Immune related (AAV & SLE) DAH.

Methods: A retrospective review of medical records of patients admitted under Rheumatology and Clinical immunology department with Immune related DAH was made with regards to their presentation, treatment & response, mortality, morbidity and long term outcome. Study was performed after approval and ethical clearance from IRB.

Results: From June 2012 to Augst 2016, 18 patients (15 were AAV related & 3 as SLE related) were admitted. Amongst AAV patients, PR3 positive were 11 & MPO positive were 4. Fourteen patients were females and 4 males, age ranged from 14 – 68 yrs (median=54.5 yrs). Mean duration of disease before onset of DAH was 3 months. Nine (50%) patients had associated kidney and musculoskeletal involvement. Eleven (61.11%) patients were admitted under ICU care requiring artificial ventilation. Pulse methylprednisolone injections were given in 15 (83.33%), Cyclophosphamide in 13 (72.22%), IVIg in 2 (11.11%), plasmapheresis in 7 (38.88%) patients. Time from first consultation to pulse methylprednisolone was in range from 1 to 5 days. Out of 18, 11 patients achieved remission. In hospital mortality was seen in 5 (27.77%) patients, all were AAV (MPO+=3, PR3+=2), all were complicated with sepsis with MODS before death. Out of 7 who received plasmapheresis, 2 patients (28.4%) died, 2 patients developed CKD (dialysis independent). Duration of ICU& hospital stay in days ranged from 3 to 28 days & 2 to 40 days respectively. Mean follow up was 16 months (range 11–42 months) on OPD visits. Two had relapse on follow up (1 nephritis, 1 persistent cavities with episcleritis) who were given Rituximab. Total 5 (38.46%) received Rituximab out of which 2 were refractory to Cyclophosphamide, 2 had relapse & 1 concomitantly. Eight patients (44.44%) developed morbidity in the form of dialysis independent-CKD in 4 (PR3+ in 2, MPO+ in 1 and Lupus nephritis in 1) with concomitant cataract in 1, ILD in 2, hearing loss in 1 and finger amputation in 1). All 13 patients who survived are in remission. Ongoing maintenance treatment is Azathioprine in 5, Mycophenolate mofetil in 3 and Rituximab in 3 patients.

Conclusions: High index of suspicion with early diagnosis and treatment results in low mortality and better long term outcome. All mortality was because of delay in diagnosis. Rituximab is effective in achieving remission in refractory as well as relapsed cases

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