FRIDAY, 16 JUNE 2017

WIN & HOT session

SP0107 HOT SESSION: SCLERODERMA TREATMENT

C.P. Denton. *Centre for Rheumatology, Royal Free Hospital and Ucl Medical School, London, United Kingdom*

This presentation will provide an up to date summary of current management of SSc that can be applied across the disease spectrum. Systemic sclerosis (SSc) remains a challenging multifaceted rheumatic disease with high mortality and morbidity. However, treatments are emerging for some aspects of the disease and long term survival has improved significantly over the past decades. This session will review the clinical challenge and current therapeutic landscape of SSc focusing on practical aspects of management such as identifying and treating significant organ based complications in the lung, heart, kidney and gastrointestinal tract. Current approaches to overall disease management will be summarized including the use of haemopoietic stem cell transplant in selected poor prognosis cases. Recently updated EULAR/EUSTAR treatment recommendations will be reviewed and other evidence based management guidelines will be considered within a practical patient-focused framework.

Disclosure of Interest: C. Denton Grant/research support from: Inventiva, CSL Behring, GSK, Bayer, Consultant for: GSK, Actelion, Inventiva, Roche

DOI: 10.1136/annrheumdis-2017-eular.2534

FRIDAY, 16 JUNE 2017

Comorbidities in rheumatoid arthritis

SP0108 HERPES ZOSTER: HOW TO PREVENT, TO DIAGNOSE AND TO TREAT

L. Calabrese. *Rheumatology, Cleveland Clinic, -Cleveland, United States*

Herpes Zoster is a major public health problem and is an infection that results from re-activation of latent varicella infection acquired most commonly naturally or more recently through immunization. The incidence of HZ is approximately twice that of the general population in patients with immune mediated inflammatory diseases (IMIDs). Underlying mechanisms are largely those which compromise cell mediated immunity and epidemiologic risks largely follow immunosenescent patterns (i.e. aging). Rheumatologists use a large variety of immunosuppressive drugs which further increase the risk of HZ and are obliged to recognize the clinical syndrome, its complications, apply effective therapy and be actively engaged in strategies to maximize immunization and prevention. This discussion will focus on recent advances in each of these areas highlighting newly described complications of HZ such as stroke and advances in vaccine development.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.7214

FRIDAY, 16 JUNE 2017

Life-threatening presentation of rheumatic diseases

SP0109 ACUTE RESPIRATORY FAILURE, MACULO-PAPULAR RASH, INDURATIVE EDEMA OF THE EXTREMITIES AND CERVICAL LYMPHADENOPATHY IN A 6-WEEK-OLD INFANT

C. Birolo. *Pediatric Rheumatology Unit, Department for Woman and Child Health, University of Padua, Italy, Padua, Italy*

Case report: A previously healthy 40-days-old male infant, from non-consanguineous parents from Morocco, presented in a peripheral hospital with fever for 48 hours, associated with rhinorrhea, mild diarrhea and progressive irritability during the last 24 hours. Initial laboratory studies revealed elevated acute phase reactants (CRP 116 mg/L), mild neutrophilia, elevated liver enzymes (AST 315, ALT 174 U/L), direct hyperbilirubinemia, discoagulopathy. Cerebral spinal fluid analysis and microbiology cultural workup resulted negative. A wide-spectrum antibiotic and antiviral therapy was initiated. On the 6th day of illness he developed diffuse maculo-papular rash, indurative edema of the extremities, right cervical lymphadenopathy and bilateral conjunctival injection. On the basis of a certain clinical diagnosis of Kawasaki disease and the infant was treated with IGIV 2 g/kg. An echocardiography performed prior to the IGIV infusion showed homogeneously dilated coronary arteries (left coronary artery 3.2 mm, right coronary artery 2.2 mm). Twelve hours after the end of IVIG infusion, the child presented a rapidly progressive, severe respiratory failure requiring endotracheal intubation and was transferred to our ICU. On admission (day 7th), physical examination revealed a feverish, critically ill-infant with hepatomegaly (5–6 cm below the right costal margin), diffuse maculo-papular rash, “sock-like” erythema and swelling of the feet, cheilitis, bilateral conjunctival injection and right cervical adenopathy. The urine output was markedly decreased; he rapidly developed hemodynamic instability with hypotension and tachycardia. Complete blood count showed anemia (6.8 g/dl), thrombocytopenia (16.000/mmc), elevated CRP (240 mg/L), hypoalbuminemia (18 g/dl) and hypofibrinogenemia (0.83 g/L); liver enzymes were normal. Intensive ventilatory and hemodynamic support therapy were started, in addition to a massive transfusional regimen.

Given the clinical and hematological picture, the diagnosis of MAS was considered and subsequently confirmed by high ferritin level (2197 mcg/L), AST above the normal value (61 U/L) and hypertriglyceridemia (181 mg/dl) [2]. The clinical suspicion was supported by persistent cytopenia despite daily transfusions, low erythrocyte sedimentation rate (3 mm/h) with concomitant rising CRP, elevated IL2-R level (28.320 KU/L) and decreased NK function. The patient was treated with high dose methylprednisolone pulse therapy (25 mg/kg) for 3 consecutive days 12–14), followed by a maintenance of 1 mg/kg/daily. By day 15th, a progressive decrease in inflammatory markers and a concomitant improvement of general conditions was observed, with the possibility to discontinue inotropic support on day 12th and invasive ventilation on day 25th. Since day 17th, a diffuse cutaneous desquamation was noted. The fever settled on day 35th.

Echocardiography follow-up revealed an increasing, irregular dilation of left (max 5 mm) and right (max 3.5 mm) coronary arteries, with a progressive left ventricular apex hypocinesis, but a stable ejection fraction (55%). ECG showed persistent repolarization abnormalities.

Of note, Adenovirus-PCR was found positive in the bronchoalveolar washing performed on admission.

The differential diagnosis included: Kawasaki disease complicated by respiratory distress syndrome and MAS, familiar hemophagocytic lymphohistiocytosis, autoimmune lymphoproliferative syndrome, criopyrinopathies and immunodeficiency.

Comment: Kawasaki disease can be catastrophic in the early infancy, due to atypical presentation and resistance to the conventional therapy. Despite typical in its presentation, this case was complicated by two challenging conditions, respiratory distress syndrome and MAS, which hampered the diagnostic and therapeutic management during the course of the disease and required massive intensive support.These two complication are rarely associated to KD. In this particular case, respiratory failure can probably be explained by a combination of causes: fluid overload, systemic vasculitis and the concomitant and probably triggering Adenovirus infection.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.7176

SP0110 LIFE-THREATENING COMPLICATIONS IN SYSTEMIC VASCULITIS

M.C. Cid on behalf of G. Espigol-Frigolé, S. Prieto-González, J. Hernández-Rodríguez, A. García-Martínez. *Systemic Autoimmune Diseases, Hospital Clinic. University of Barcelona, Idibaps, Barcelona, Spain*

Systemic vasculitis may present with life-threatening complications that need to be promptly recognized and appropriately managed to ensure patient survival and minimize irreversible organ damage.

The most common life-threatening events differ between large and medium or small-vessel vasculitis. In large-vessel vasculitis, particularly giant-cell artertis (GCA), vascular remodelling in response to inflammation may lead to severe stenosis leading to ischemic stroke in 3–6% of patients and, rarely, myocardial infarction, mesenteric ischemia or critical extremity ischemia. For these complications, particularly when happening in treated patients, intensifying immunosuppressive therapy is not the best or only option and additional interventions may be required. Stroke occurs as a consequence of carotid or vertebral stenosis. Stenosis of the carotid siphon has been repeatedly reported. In necropsy studies, vasculitic involvement and thrombosis of proximal intracranial branches has been observed. Infarcts are usually multiple, usually happen shortly after the initiation of glucocorticoid therapy, convey a 30% mortality or lead to remarkable disability. If critical stenosis is suspected before irreversible infarction, percutaneous intraluminal angioplasty may be function and life saving.

Acute aortic syndrome (aortic dissection or intramural haematoma) is an increasingly recognized hurdle in patients with GCA. It is usually a delayed complication occurring months and frequently years after diagnosis. Its frequency has not been delimitated but in a recent prospective follow-up study it was demonstrated to affect at least 2% of patients. Emergency open surgery repair, when feasible, is the best option for ascending aorta involvement (type A) and endovascular repair for involvement of the descending aorta (type B).

In small-vessel vasculitis, life-threatening presentations include rapidly progressive glomerulonephritis leading to kidney failure, alveolar haemorrhage, alithiasic colecistitis with perforation and intestinal ischemia. Rapidly progressive glomerulonephritis and alveolar haemorrhage are more frequently seen in anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) and anti glomerular basement membrane disease. Alveolar haemorrhage can be occasionally seen in cryoglobulinemic or IgA vasculitis. In addition to supportive measures, these patients are usually treated with high-dose methyl-prednisolone, cyclophosphamide or rituximab and plasma exchange. Plasma exchange has been found superior to IV methyl prednisolone mega-doses in preserving or recovering renal function but this advantage do not seem to persist over long-term follow-up. This approach is usually applied also to alveolar haemorrhage although there is not strong evidence supporting it.

Gastrointestinal complications, particularly intestinal ischemia and intestinal or gallbladder perforation are life-threatening complications which may require emer-

gent surgery. These may be observed in polyarteritis nodosa, cryoglobulinemic vasculitis, eosinophilic granulomatosis with polyangiitis and other AAV. High-dose glucocorticoids and cyclophosphamide are usually applied.Children with IgA vasculitis may develop bowel intussusception
Deep venous thrombosis and pulmonary embolisms are significantly more frequent in AAV and Behçet disease than in the general population, especially during active disease. Anti-coagulation may be needed in AAV, although this approach is controversial in Behçet's disease. By contrast, aneurysm formation is typical in polyarteritis nodosa and Behçet's disease and may be occasionally seen in AAV. Massive bleeding derived form aneurysm rupture usually requires arterial embolization.
It is important to keep in mind that during the early course of diagnosed vasculitis, intense immunosuppressive therapy may favour life-threatening infections including opportunistic infections such as pneumocystis jiroveci pneumonia or disseminated CMV.
In summary, systemic vasculitis may present with a variety of severe complications and other may develop during follow-up. These complications are heterogeneous, vary according to the size of vessels involved, and usually require specific procedures or treatments in addition to immunosuppressive therapy. Due to the life-threatening nature of these complications their immediate recognition and management are crucial to patient survival.
Supported by SAF 577–8R and Marató TV3 2014/201507
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7166

FRIDAY, 16 JUNE 2017

AxSpA: From bug to gut and to disease phenotype –

SP0111 INHIBITING BONE FORMATION IN THE CLINIC. ARE WE THERE YET?

R. Landewe. *Rheumatology& immunology, Amsterdam Rheumatology & immunology Center, Amsterdam, Netherlands*

One of the most characteristic features of axial spondyloarthritis (axSpA) is bone formation in the spine (syndesmophytes). Syndesmophytes may occur at any time during the course of the disease, are more frequent in patients with radiographic axSpA (AS) than in those with non-radiographic axSpA, and are best seen on conventional X-rays of the spine. Currently, it is suggested that (low-radiation) CT-scanning of the spine provides a better (more sensitive) picture of developing syndesmophytes than conventional X-rays.
Syndesmophytes matter in that they interfere with spinal mobility and physical function independent of inflammation. As such, it makes sens to try and prevent their occurrence or to inhibit their progression.
It is a matter of debate whether current available treatments are able to inhibit syndesmophyte growth or occurrence. Part of the debate is the methodological challenges related to measuring syndesmophyte progression properly.
In this lecture I will address current issues related to inhibition of syndesmophyte formation in patients with axial SpA.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7211

FRIDAY, 16 JUNE 2017

Pregnancy meets rheumatic patients

SP0112 WHICH DRUGS IN PREGNANT PATIENTS?

M. Ostensen. *Department of Rheumatology, St.Olavs Hospital, Trondheim, Norway*

Management of rheumatic disease during pregnancy starts with prepregnancy counselling.Assessment of maternal and fetal risks is necessary for adjusting therapy before and during pregnancy. The aim of therapy is to keep the disease in remission or at least at low activity throughout pregnancy.
Immunosuppressive drugs requiring withdrawal before conception are methotrexate, cyclophosphamide,and mycophenolate which are known teratogenic drugs. Other drugs like leflunomide, tofacitinib and several biological should be discontinued because pregnancy experience is at present insufficient and safety for the fetus has not been proven.Flares of rheumatic disease showing be treated immediately and with pregnancy compatible drugs.For patients with inflammatory arthritis like rheumatoid arthritis,spondyloarthritis and juvenile idioipathic arthritis disease activity during pregnancy can be controlled with antimalarials, sulfasalazine and TNF inhibitors. Women with systemic lupus erythematosus should continue basic therapy with hydroxychloroquine, and azathioprine, ciclosporine or tacrolimus added when necessary due to organ manifestations. Severe flares during pregnancy may require biologics like rituximab,abatacept,tocilizumab or Anakonda, in SLE corticoid pulses or, if life threatening, intravenous gamma globulin or cyclophosphamide.
Treatment during pregnancy demands balancing suppression of maternal disease and no harm to the child. Selecting the adequate type, the right dose and the right timing of medications for optimal care of pregnant patients remains a challenge.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7246

SP0113 PREGNANCY IN SLE: STILL CHALLENGING FETAL AND MATERNAL ISSUES

R. Fischer-Betz. *Rheumatology, Heinrich-Heine-University, Duesseldorf, Germany*

Patients with SLE are mostly young women diagnosed during their childbearing years. Several "unmet needs" in the management of reproductive health issues may impact on the decision to have children. Because of earlier recognition of disease and advances in medical treatment, family planning has gained greater importance. Concerns include the effect of pregnancy on maternal disease, the impact of disease activity on fetal health, and the safety of medications during pregnancy and breastfeeding. Preconception counselling and risk stratification (including life style, disease activity, autoantibody profile, previous vascular and pregnancy morbidity, hypertension and the use of drugs with emphasis on benefits from hydroxychloroquine and antiplatelets/anticoagulants) are essential for prevention of unwanted complications during pregnancy. Recommendations for the management of family planning and antirheumatic treatment during pregnancy and lactation have been published recently by EULAR. However, many lupus patients still do not feel that their family planning concerns are adequately addressed in current clinical practice and report that they receive inconsistent advice from the various healthcare professionals. There is a clear need for provision of up-to-date and consistent information/support to our patients.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7091

SP0114 CHILDREN OF PATIENTS WITH RHEUMATIC DISEASES: ISSUES RELATED TO MATERNAL DISEASE AND TREATMENT

L. Andreoli. *Rheumatology and Clinical Immunology, University of Brescia, Brescia, Italy*

A major source of anxiety for women with systemic autoimmune diseases (SADs) who wish to become pregnant is the possible impact of maternal disease and medications on the offspring, in terms of physical and mental development. A recent multicenter survey conducted in 24 Italian Rheumatology Centers showed that more than 50% women affected by SADs restricted their family size mainly because they were afraid that children could get an autoimmune disease or could be harmed by intrauterine exposure to maternal autoantibodies and anti-rheumatic drugs (Dall'Ara, ACR abstract, Arthritis Rheumatol 2016; 68, suppl 10). Therefore, the long-term follow-up children born to mothers with SADs is a topic of major relevance for the counselling on family planning.
First of all, it should be emphasized that preterm birth and other foetal complications, such as low birth weight and babies small for gestational age, are more common in patients with systemic autoimmune diseases as compared to the general population. These conditions carry themselves an increased risk for poorer physical and neuropsychiatric development. Therefore, the prevention of foetal complications should be operated by means of close obstetrical monitoring and tight control of maternal disease activity, which would be detrimental for foetal wellbeing. In this context, the use of "safe" anti-rheumatic drugs is of paramount importance for pregnant women with SADs.
Recently, a dedicated EULAR Task Force has released points to consider for the use of anti-rheumatic drugs during pregnancy and lactation (Gotestam Skorpen, Ann Rheum Dis 2016). The work of this Task Force was focused on updating the information about the use of "conventional synthetic" (cs) DMARDs but also to provide for the first time evidence-base indications on the use of "biologic" (b) DMARDs, mainly anti-TNFalfa agents.
No significant impairment in the maturation and functioning of the child's immune system has been observed for several csDMARDs, supporting their safety of use during pregnancy (Andreoli, J Autoimm 2012).
Turning to bDMARDs, a case-control study on the long-term follow-up of children exposed in utero to anti-TNFalfa drugs showed the safety of use either until the positive pregnancy index or during the second and third trimester of gestation (Dall'Ara, EULAR abstract, Ann Rheum Dis 2016; 75, Suppl 2:493). No differences between exposed and non-exposed children were found in terms of congenital defects, developmental milestones, response to vaccinations and major health problems. No particular problems were also observed in children who were breastfed while maternal anti-TNFalfa intake. The use of anti-TNFalfa agents during breastfeeding had been proposed to women who were strongly motivated based on the following considerations: 1) these drugs are poorly or absolutely not excreted into breast milk as recently demonstrated for certolizumab pegol (Clowse, ACR abstract, Arthritis Rheumatol 2016; 68, suppl 10); 2) even this was the case, the drug will be degraded in the baby's gastrointestinal tract and absorption could not be possible.
Regarding maternal disease, major concerns are linked to fetal exposure to maternal autoantibodies, mainly anti-Ro/SSA (for the development of Neonatal Lupus) and antiphospholipid antibodies (aPL). Therefore, the evaluation of these autoantibodies with potential negative impact on pregnancy and neonatal outcome should be part of the preconception work-up of women with SADs in order to provide adequate counselling and preventative strategies (Andreoli, Ann Rheum Dis 2017).