FRIDAY, 16 JUNE 2017

WIN & HOT session

SP0107 HOT SESSION: SCLERODERMA TREATMENT

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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.

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Comorbidities in rheumatoid arthritis

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

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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

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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

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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.

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SP0110 LIFE-THREATENING COMPLICATIONS IN SYSTEMIC VASCULITIS

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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-