SAT0252 **LONG-TERM SURVIVAL ANALYSIS AND PROGNOSTIC OUTCOME FACTORS IN IDIOPATHIC INFLAMMATORY MYOPATHY**

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Background: Idiopathic inflammatory myopathy is a group of systemic connective tissue diseases, including dermatomyositis (DM), polymyositis (PM) and inclusion body myositis (IBM). PM/DM are characterized by chronic muscle inflammation and can affect other internal organs. Survival rate and prognostic factors of PM/DM vary widely. The 5-year survival rates for PM/DM range from 60% to 95%. Additionally, the long-term outcome of a large group of Chinese patients with PM/DM is absent. Although Interstitial lung disease (ILD) and coexistent cancer were indicated to be predictors of mortality, few studies conducted multivariate regression analysis to provide independent prognostic factors, as well as the calculation of Standardized Mortality Ratio for PM/DM patients.
Objectives: To analyze the survival rate, cause of death and the prognostic outcome factors in idiopathic inflammatory myopathy (IIM).
Methods: A total of 201 IIM patients treated in Peking University People's Hospital from January 1996 to September 2013 were included. Medical records were abstracted for clinical, laboratory and therapeutic data.
Results: The follow-up time of 201 IIM patients, including 121 with DM, 33 with PM and 47 with CADM, ranged from one to 492 months, and 56 cases were lost to follow up. Overall cumulative survival rate was 93.0%, 87.0%, 86.0% and 84.0% at 1, 3, 5 and 10 years respectively from the disease onset. The rates were similar in DM, PM and CADM. During the follow-up period, 26 (19.3%) patients died, among which 21 cases died within 3 years from the onset. Respiratory failure associated with ILD was the main cause of death, followed by cardiac involvement and coexistent malignancy. Rapidly progressive ILD, myalgia, fever, abnormal ECG, normal CKMB, lymphopenia and hypoalbuminemia, elevated LDH and CEA were associated with mortality ($P<0.05$). In multivariate Cox regression analysis, elevated CEA was the significant predictor of poor outcome. Therapy with glucocorticoids combined with immunosuppressants for more than one year was negatively correlated with mortality, as well as methylprednisolone pulse therapy for patients with rapidly progressive ILD. Intravenous cyclophosphamide therapy was associated with a better survival than MTX.
Conclusions: The ten-year survival rate shows no statistically significant difference among DM, PM and CADM. The survival rate of patients with rapidly progressive ILD is significantly lower than patients with chronic or without ILD. Elevated CEA acts as the independent risk factor for poor prognosis. Glucocorticoids combined with immunosuppressants for more than one year is associated with high survival rate. Intravenous cyclophosphamide therapy is associated with a better survival than MTX.
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Paediatric rheumatology

SAT0253 **EVANS SYNDROME AT CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS DIAGNOSIS: A LARGE MULTICENTER STUDY**

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Background: Evans syndrome (ES) is an uncommon manifestation characterized by autoimmune destruction of red cells and platelets and concomitant or sequential appearance of immune thrombocytopenia (ITP) and autoimmune hemolytic anemia (AIHA). This involvement has been associated to severe disease activity in adult patients with systemic lupus erythematosus (SLE), particularly at disease onset. However, ES studies in childhood-onset SLE (cSLE) patients have been rarely reported and limited to small populations.
Objectives: The objective of the present multicenter study was to assess ES in a large cSLE cohort at diagnosis evaluating prevalence, clinical features, laboratory findings and outcomes.
Methods: A retrospective multicenter cohort study (Brazilian cSLE group) was performed in 10 Pediatric Rheumatology services including 850 patients with cSLE (ACR criteria). None of them had secondary etiologies of autoimmune cytopenias, such as infections, primary immunodeficiencies and malignancies. Patients were divided in two groups for the assessment of lupus manifestations, laboratory exams and treatment at cSLE diagnosis: patients with ES and patients without ES.
Results: ES was observed in 11/850 (1.3%) cSLE patients at diagnosis. The majority of them had hemorrhagic manifestations (58%) and active disease (82%). All patients with ES were hospitalized and none of them died. Comparisons of cSLE patients with and without ES at diagnosis revealed similar frequencies of female gender, multi-organ involvement, autoantibody profile and low complement levels ($p>0.05$). Patients with ES had a lower frequency of malar rash (9% vs. 53%, p=0.003) and musculoskeletal involvement (18% vs. 69%, p=0.001) than those without this complication. The median of hemoglobin [7.4 (5.4–9.4) vs. 10.3 (3.5–16.4)g/dL, $p<0.001$] and platelets [27 (15–54) vs. 231 (2–761)x10^3/mm^3, p=0.005] were significantly lower in ES compared to non-ES patients, whereas lymphocytes were significantly higher in ES patients [1.8 (1–2.38) vs. 1.16 (0.07–7)x10^3/mm^3, $p<0.001$]. The frequencies of intravenous methylprednisolone (82% vs. 43%, p=0.013) and intravenous immunoglobulin use (64% vs. 3%, $p<0.0001$) were significantly higher in the former group. Current prednisone dose between the two groups was similar [1.1 (0.76–1.5) vs. 1.0 mg/kg/day (0–30), 0.195].
Conclusions: Our large multicenter study identified that ES was a rare and severe cSLE manifestation with a difficult diagnosis due to the absence of typical lupus manifestations, often requiring hospitalization and intravenous treatment.
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SAT0254 **THE ICEBERG IN JUVENILE ONSET SYSTEMIC LUPUS ERYTHEMATOSUS: SUBCLINICAL DETERIORATION OF CARDIAC FUNCTIONS ASSESSED WITH TWO-DIMENSIONAL SPECKLE TRACKING ECHOCARDIOGRAPHY AND CONTRIBUTING FACTORS OF SYSTOLIC DYSFUNCTION**

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Background: Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by vasculitis and inflammation in various organs. Cardiovascular involvement, although not frequent in juvenile-onset SLE (j-SLE), if present, is a significant cause of morbidity and mortality (1). Particularly, involvement of the myocardium layer may lead to ventricular dysfunction since its progression is insidious (1,2). Speckle tracking echocardiography (STE) could demonstrate subclinical myocardial deformations (strain) that globally seems normal in conventional echocardiography (2).
Objectives: The aim of this study is early detection of subclinical systolic dysfunctions in j-SLE with STE and if present, then to investigate whether this is disease-related or it is a result of other predisposing conditions.
Methods: 35 patients with j-SLE and 30 healthy children as a control group were evaluated between January and August 2015 at outpatient clinics of Cerrahpasa Medical Faculty. STE was performed on all patients and controls. Medical records that are including age at diagnosis, duration of the disease, diagnostic criteria, laboratory tests and cumulative clinical manifestations were evaluated. SLE disease activity was assessed using the SLE Disease Activity Index (SLEDAI). A SLEDAI score >4 was arbitrarily designated as a sign of moderate/severe disease activity.
Results: j-SLE patients had lower EF values than control subjects. Left ventricular end diastolic dimension (LVEDD) and left ventricular end systolic dimension (LVESD) were significantly greater in j-SLE patients (42,278±4,530 vs. 37,314±5,535; 28,108±3,344 vs. 24,055±3,290 p=0.001, respectively) than in the control group. There was a significant reduction in systolic parameters of longitudinal strain in the j-SLE group ($p<0.05$) at all segments compared to control patients.
SLE patients were divided into two subgroups. Group 1 included patients having SLEDAI scores >8 at the beginning of the disease but who improved with therapy during follow up, with resulting SLEDAI scores less than or equal to 4 points. Group 2 included j-SLE patients with SLEDAI scores >8 at diagnosis but with SLEDAI scores still greater than 4 at the end of follow up.
In comparisons of two groups, mid inferior and mid inferolateral LV segment STE